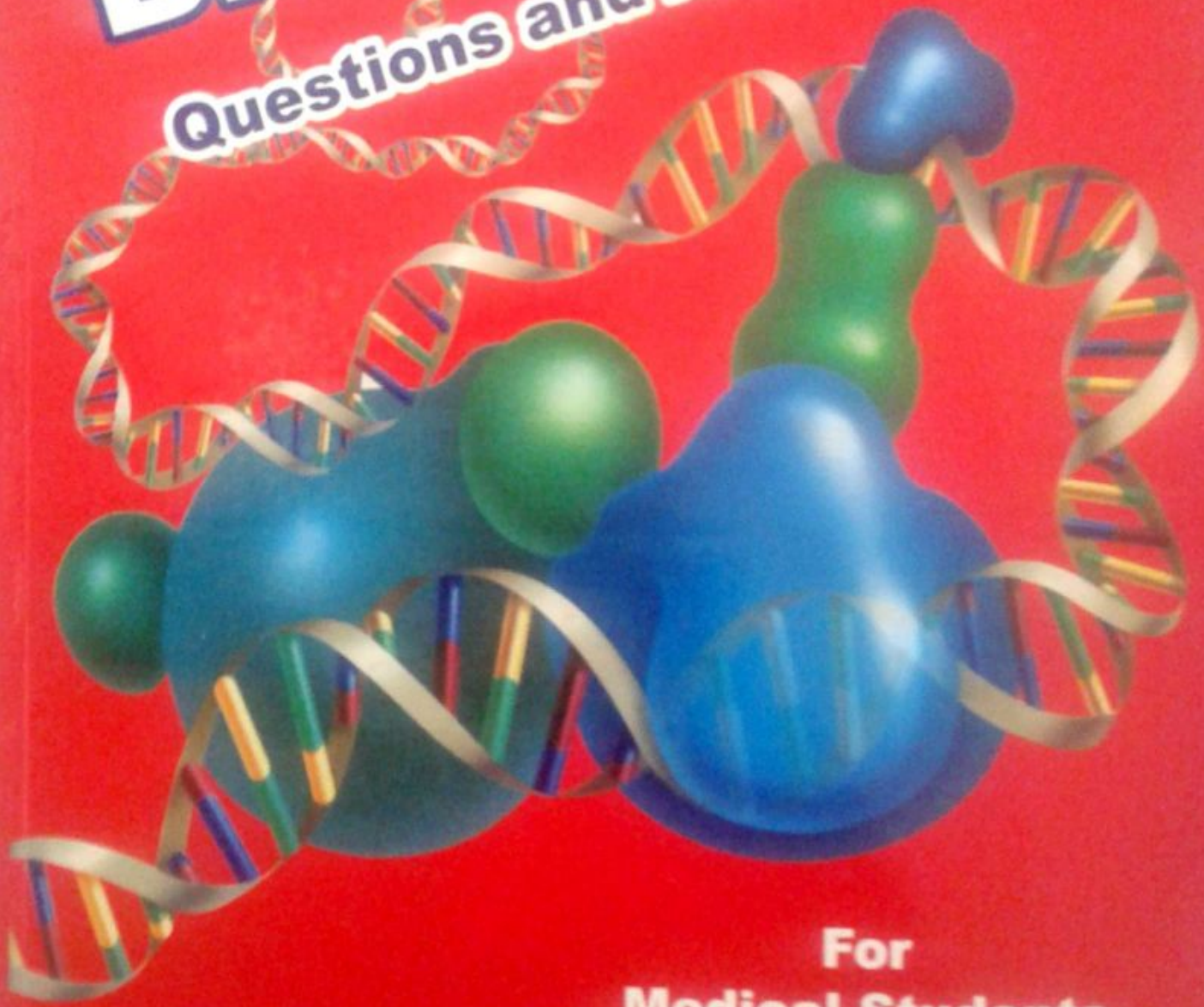
Oraby's
Illustrated
Reviews of

Biochemistry

Questions and Answers



For
Medical Students
and Postgraduates
Part IV (Volume 2)

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Chapter 14 *Carbohydrate Metabolism*

1. What is the end product of action of pancreatic amylase on starch?

   A. Maltose.

2. *Which is more effective salivary amylase or pancreatic amylase?*

   A. Pancreatic amylase.

3. *Why?*

   A. Because salivary amylase is rapidly inactivated by gastric HCl

4. *Cellulose and starch are polysaccharides made of glucose, but cellulose cannot be digested by human beings, why?*

   A. Cellulose contains β 1,4 linkages, which cannot be digested by human enzymes.

5. *What is lactose intolerance?*

   A. This is a deficiency of lactase enzyme, which digests lactose into glucose and galactose.

6. *What are effects of lactose intolerance?*

   A. Dehydration, diarrhea distention, and abdominal cramps.

7. *What are mechanisms of these effects?*

   A. The presence of lactose in intestine causes increased osmotic pressure: So water will be drawn from the tissue (causing dehydration) into the large intestine (causing diarrhea). It also causes increased fermentation of lactose by intestinal bacteria that ferment lactose with subsequent production of CO_2 gas. This causes distention and abdominal cramps.

8. *The rate of absorption of sugars in intestine is highest for which monosaccharide?*

   A. Absorption rate of galactose is more than glucose, while fructose is absorbed at a lesser rate than glucose.

9. *How glucose is absorbed from intestine?*

A. Through active transport by carrier protein named sodium dependent glucose transporter (SGluT).

10. *How glucose is released from intestinal cells into the blood stream?*

A. By glucose transporter type 2 (GluT2).

11. *How glucose is taken up by cells from blood stream?*

A. In tissues GluT2 is involved in absorption of glucose from blood.

12. What is the importance of GluT4?

A. It is the glucose transporter present in muscle and adipose tissues. Insulin induces synthesis of these transporters. In diabetes mellitus, entry of glucose into muscle is decreased, because GluT4 is reduced in insulin deficiency.

13. *What is glycolysis?*

A. It is oxidation of glucose to give pyruvate (in the presence of oxygen) or lactate (in the absence of oxygen), along with production of a small quantity of energy.

14. *What is the net yield of ATP from one molecule of glucose in anaerobic glycolysis?*

A. 2 ATP.

15. *What is the net yield of ATP from one molecule of glucose in aerobic glycolysis?*

A. 6 or 8 ATP.

16. *When aerobic glycolysis gives 6 or 8 ATPs?*

A. 6 ATP if 2 $NADH+H^+$ transfers its hydrogen to mitochondria through glycerophosphate shuttle and 8 ATP if 2 $NADH+H^+$ transfers its hydrogen to mitochondria through aspartate malate shuttle.

17. *In which condition pyruvate is produced, and when lactate?*

A. In aerobic condition, pyruvate is produced. When oxygen is lacking, lactate is produced.

18. *What is the significance of glycolysis?*

A. It is the only pathway that is taking place in all the cells of the body. Glycolysis is the only source of energy in RBCs. Moreover,

anaerobic glycolysis forms the major source of energy in actively contracting muscles.

19. *What are other functions of glycolysis?*

A. Apart of energy production, glycolysis helps the oxygenation of tissues through formation of 2,3 bisphosphoglycerate which decreases the affinity of hemoglobin to O_2 and provides important intermediates.

20. *What is the function of 2,3-bisphospho glycerate?*

A. When combined with hemoglobin, 2,3-BPG reduces the affinity towards oxygen. This leads to good oxygen delivery to tissues.

21. *What are important intermediates produced by glycolysis?*

A. Dihydroxyacetone phosphate used for lipogenesis, 3-phosphoglycerate used for synthesis of serine, pyruvate used for synthesis of alanine and in the presence of O_2 it can pass into mitochondria → acetyl CoA → Krebs' cycle.

22. *Which tissues prefer anaerobic glycolysis?*

A. Mainly RBCs, and exercising muscle.

23. *Why RBCs are very dependent upon glycolysis for ATP production?*

A. Because mature RBCs contain no mitochondria.

24. *Why exercising muscles are dependent upon glycolysis for ATP production?*

A. Because they undergo frequent oxygen lack.

25. *Which other tissues prefer anaerobic glycolysis?*

A. Tissues with no mitochondria as cornea and lens. Tissues with few mitochondria as testis, leucocytes, medulla of the kidney, retina, skin and gastrointestinal tract and cancer cells as they undergo frequent oxygen lack.

26. *What is the importance of phosphofructokinase?*

A. It is one of the key enzymes (rate limiting enzymes) of the glycolysis. It is an irreversible reaction.

27. *What is the substrate for aldolase A reaction?*

A. Fructose-1,6-bisphosphate.

28. *During glycolysis, energy is produced during which steps?*

A. (1) Step 1,3-bisphosphoglycerate to 3-phospho glycerate, (2) step, phosphoenol pyruvate to pyruvate.

29. *Fluoride ions inhibit which enzyme?*

A. Enolase.

30. *What is the importance of the above inhibition?*

A. Fluoride is used to prevent glycolysis, as preservative for blood before glucose estimation.

31. *NAD is reduced to NADH in which reaction of glycolysis?*

A. Glyceraldehyde-3-phosphate dehydrogenase reaction.

32. *How NAD^+ can be regenerated from $NADH+H^+$ in glycolysis?*

A. This can be done by oxygen, but when oxygen is lacking this can be done by conversion of pyruvate to lactate (last reaction of glycolysis).

33. *What is the purpose of lactic acid production under anaerobic conditions?*

A. For regeneration of NAD^+. This helps continuity of glycolysis, as the generated NAD^+ will be used once more for oxidation of another glucose molecule.

34. *As the end product of glycolysis, pyruvate and NADH are formed. During anaerobic conditions, this NADH is reconverted to NAD+ by what mechanism?*

A. Lactate dehydrogenase reaction.

35. *As the end product of glycolysis, pyruvate and NADH are formed. During aerobic conditions, this NADH is reconverted to NAD+ by what mechanism?*

A. Oxygen, in respiratory chain reactions.

36. *What is Cori's cycle?*

A. It is the conversion of glucose into lactate in muscle, followed by conversion of lactate into glucose in liver. It is then taken up through gluconeogenesis pathway, and becomes glucose. This glucose can enter into blood and then taken to muscle.

37. *What is the other name of Cori cycle?*

A. Lactic acid cycle.

38. *What is the purpose of Cori's cycle?*

A. By this means, the lactate is efficiently reutilized by the body.

39. *Why lactate is transported from muscle to liver?*

A. Oxygen is limited in exercising muscles, so lactic acid could not be transformed to pyruvate in muscles. It is transported to liver, where it is transformed to pyruvate and then to glucose.

40. *What are key glycolytic enzymes?*

A. Glucokinase, hexokinase, phosphofructokinase, and pyruvate kinase.

41. *What is hexokinase?*

A. Hexokinase is the first enzyme in the glycolysis. It phosphorylates glucose to glucose-6-phosphate. It is present almost in all tissues.

42. *What is glucokinase?*

A. Glucokinase - together with hexokinase - are the first enzymes in the glycolysis. However, glucokinase is present only in liver. It acts only on glucose. It is most active when glucose level in blood is increased after meals.

43. *What are mechanisms of regulation of glycolysis?*

A. Hormonal regulation, energy regulation and substrates regulation.

44. *What are hormones regulating glycolysis?*

A. Insulin and glucagon.

45. *How can insulin regulate glycolysis?*

A. Insulin stimulates glycolysis. It stimulates synthesis of all key enzymes of glycolysis (except hexokinase).

46. *When insulin is secreted?*

A. It is secreted after meal (in response to high blood glucose level).

47. *How can glucagon regulate glycolysis?*

A. It inhibits the activity of the key enzymes of glycolysis.

48. *When glucagon is secreted?*

A. It is secreted in response to low blood glucose level.

49. *What is energy regulation of glycolysis?*

A. High level of ATP inhibits both PFK-1 and pyruvate kinase. High levels of ADP and AMP stimulate PFK-1.

50. *What are substrates regulating glycolysis?*

A. Glucose-6-phosphate, fructose 1,6 bisphosphate fructose 2,6 bisphosphate, and citrate.

51. *What are the inhibitors of phosphofructokinase?*

A. ATP, citrate, and glucagon.

52. *What are the activators of phosphofructokinase?*

A. AMP, Fructose-2,6-bisphosphate, and insulin.

53. *What are substrates that regulate glycolysis?*

A. Glucose-6-phosphate inhibits hexokinase, fructose 1,6 bisphosphate stimulates pyruvate kinase, fructose 2,6 bisphosphate stimulates phosphor-fructokinase-1 (glycolysis) and inhibits fructose 1,6 bisphosphatase (gluconeogenesis), and citrate inhibits phosphofructokinase-1.

54. *Malate-aspartate shuttle is used for what purpose?*

A. (1) It helps the transfer of hydrogen carried by $NADH+H^+$ - produced in the course of glycolysis in hepatic cells- to inside the mitochondria. (2) Reactions of gluconeogenesis are taking place in cytosol. Hence, the oxaloacetate has to be transported from mitochondria to cytosol. The malate shuttle achieves this.

55. *What is the effect of deficiency of pyruvate kinase (PK) enzyme?*

A. This leads to excessive hemolysis of RBCs → hemolytic anemia.

56. *Why deficiency of pyruvate kinase (PK) enzyme causes anemia?*

A. Genetic deficiency of PK enzyme → ↓ rate of glycolysis and decrease production of ATP. ATP is required for Na^+ - K^+ ATPase, which is important for stability of RBCs.

57. *What is the effect of deficiency of hexokinase enzyme?*

A. This leads to hemolytic anemia due to decrease ATP production. The mechanism is similar to that of pyruvate kinase deficiency.

58. *What are sources of lactate?*

A. From glycolysis especially in RBCs (due to absence of mitochondria) and muscle during exercises (due to oxygen lack).

59. *What is fate of lactate?*

A. Glucose formation through Cori cycle, conversion into pyruvate (If oxygen gets available), accumulation in muscles causing muscle fatigue and finally excretion in urine and sweat.

60. *What is substrate level phosphorylation?*

A. It is a phosphorylation of ADP to form ATP at the level of the reaction itself.

61. *Give example of substrate level phosphorylation in glycolysis?*

A. Step 1,3-bisphosphoglycerate to 3-phospho glycerate, and step, phosphoenol pyruvate to pyruvate.

62. *Why lactate is the end product of glycolysis in RBCs?*

A. Because mature RBCs contain no mitochondria.

63. *Do RBCs undergo no glycolysis in diabetes mellitus?*

A. Glycolysis proceeds normally in RBCS. This is because glucose uptake by RBCs is independent on insulin hormone.

64. *What is the relationship between glycolysis and methemoglobin in RBCs?*

A. Glycolysis produces $NADH,H^+$ which is used for reduction of met-hemoglobin into hemoglobin in red cells. This reaction is catalyzed by cytochrome b_5-met-hemoglobin reductase system (cyt b_5).

65. *What is lactic acidosis?*

A. It is a metabolic acidosis resulting from increased formation of lactate or decreased utilization of lactate.

66. *What are the causes of decreased utilization of lactate?*

A. Tissue anoxia or lack of oxygen as in myocardial infarction. This is because oxygen is essential for conversion of lactate into pyruvate, which proceeds into acetyl CoA→Krebs' cycle.

67. *What is fermentation?*

A. It is the conversion of glucose into CO_2 ethanol by yeast enzymes.

68. *What is pyruvate kinase?*

A. It catalyses the reaction,phospho enol pyruvate to pyruvate.

69. *What is the effect of pyruvate kinase enzyme deficiency?*

A. Hemolytic anemia

70. *What is catalytic activity of lactate dehydrogenase (LD)?*

A. Conversion of pyruvate to lactate. The reaction is reversible.

71. *What is the medical importance of lactate dehydrogenase?*

A. This enzyme has 5 isoenzymes (LDI). Serum LDI 1 increases in certain heart diseases (myocardial infarction). Serum LDI 5 increases in certain liver diseases (infective hepatitis).

72. *What are in vitro inhibitors of glycolysis?*

A. Arsenate, iodoacetate and fluoride ions.

73. *What are the mechanisms of inhibition of each?*

A. Arsenate competes with inorganic phosphate (Pi) in the reaction catalyzed glycerladhyde-3-phosphate dehydrogenase, iodoacetate inhibits glycerladhyde-3-phosphate dehydrogenase and fluoride ions inhibits enolase.

74. *How can extramitochondrial $NADH+H^+$ be oxidized?*

A. Through Glycerophosphate shuttle and aspartate malate shuttle.

75. *Is glycolysis pathway reversible?*

A. Yes except 3 reactions (those catalyzed by kinase enzymes), Glucose-6-P → Glucose, Fructose 1,6 diphosphate → fructose-6-P and Pyruvate → Phosphoenol pyruvate.

76. *What is oxidative decarboxylation of pyruvate?*

A. It is the conversion of pyruvate into acetyl CoA.

77. *Pyruvate is converted to acetyl CoA by which enzyme?*

A. Pyruvate dehydrogenase.

78. *What are the co-enzymes necessary for oxidative decarboxylation of pyruvate?*

A. Thiamine pyrophosphate, NAD, FAD, Lipoic acid, and Co-enzyme A.

79. *How can pyruvate dehydrogenase be regulated?*

A. The enzyme is present in 2 forms active dephosphorylated and inactive phosphorylated. Factors stimulating the enzyme including insulin, pyruvate Co-enzyme A and NAD^+. Factors inhibiting the enzyme include acetyl CoA, $NADH+H^+$, ATP and calcium ions.

80. *What is citric acid cycle(TCA)?*

A. TCA is a series of reactions in which acetyl CoA is oxidized into CO_2, H_2O and energy.

81. *Which is the amphibolic pathway?*

A. Citric acid cycle because it has anabolic and catabolic functions.

82. *What are functions of Krebs' cycle (TCA)?*

A. Production of energy (12 ATP), catabolic functions and anabolic functions.

83. *What are the catabolic functions of Krebs' cycle (TCA)?*

A. TCA is the final common pathway for oxidation of carbohydrates (glucose), lipids (fatty acids) and proteins (amino acids).

84. *What are the anabolic functions of Krebs' cycle (TCA)?*

A. Synthesis of amino acids, glucose, heme, fatty acid, cholesterol and CO_2.

85. *How many ATPs are generated per one rotation of the citric acid cycle?*

A. 12 ATP.

86. *What is energy (ATP) production of complete oxidation of pyruvate?*

A. 15 ATP

87. *In hepatic cell and during complete oxidation, what is the net yield of ATP from one glucose molecule?*

A. 38 ATP.

88. *Acetyl CoA is produced from what substrates?*

A. Carbohydrate (Pyruvate), fatty acids, and leucine amino acid.

89. *Acetyl CoA is used for what purposes?*

A. Oxidation in TCA cycle, fatty acid synthesis, cholesterol synthesis, and ketone bodies formation.

90. *What is energy (ATP) production of complete oxidation of Acetyl CoA?*

A. 12 ATP

91. *Which is the substrate level phosphorylation step in the TCA cycle?*

A. Succinate thiokinase.

92. *What is the inhibitor of succinate dehydrogenase?*

A. Malonate.

93. *What are other in vitro inhibitors of citric acid cycle (CAC)?*

A. Flouroacetate, which inhibits citrate synthase and arsenate, which inhibits α–ketoglutarate.

94. *Give examples of reactions in which NADH is generated.*

A. Pyruvate dehydrogenase, isocitrate dehydrogenase, α-keto glutarate dehydrogenase and malate dehydrogenase.

95. *What are sources of oxaloacetate?*

A. Oxidation of malate, transamination of aspartate (by AST), carboxylation of pyruvate, cleavage of citrate (by citrate lyase)

96. *What are fate of oxaloacetate?*

A. Formation of citrate (by citrate synthase), reduction to malate(by malate dehydrogenase) and transamination into aspartic acid (by AST).

97. *What are sources of succinyl CoA?*

A. Oxidation of odd number fatty acids, citric acid cycle and catabolism of isoleucine, valine and methionine amino acids.

98. *What are fate of succinyl CoA?*

A. Glucose synthesis (gluconeogenesis), heme synthesis, oxidation in citric acid cycle, activation of ketone bodies and detoxication.

99. Give example of substrate level phosphorylation in Krebs' cycle?

A. The step succinyl CoA to succinate catalyzed by succinate thiokinase enzyme.

100. What is Pasteur effect?

A. It is the inhibition of glycolysis (anaerobic oxidation) by the presence of oxygen.

101. What are the steps in which carbon dioxide is liberated, during the oxidation of glucose?

A. Pyruvate dehydrogenase, isocitrate dehydrogenase, α-ketoglutarate dehydrogenase.

102. Give examples of α-ketoacids as a source of CO_2:

A. Pyruvic acid → (Acetyl CoA + CO_2), α-Ketoglutarate (TCA) → (Succinyl CoA + CO_2), and oxalo-succinate (TCA) → (α-ketoglutrate + CO_2).

103. Give examples of amino acid decarboxylation as a source of CO_2:

A. Glutamic acid → GABA + CO_2, histidine → histamine + CO_2 and tyrosine → dopamine + CO_2

104. What are sources of CO_2?

A. α-Ketoacids, decarboxylation of amino acids, fermentation, pyrimidine catabolism and pentose phosphate pathway.

105. What are fate of CO_2?

A. Carboxylation of the following substrates: pyruvate to Oxaloacetate (gluconeogenesis), acetyl CoA to malonyl CoA (fatty acid synthesis), ammonia, ATP to carbamoyl phosphate (synthesis of urea and pyrimidine), formation of C_6 of purine and synthesis of bicarbonate buffer.

106. What are vitamins needed for CAC?

A. Vitamin B2, niacin, thiamin (B_1) and pantothenic acid.

107. What is the role of vitamin B2 in CAC?

A. formation of FAD, it is the coenzyme for α-ketoglutarate dehydrogenase complex and succinate dehydrogenase.

108. What is the role of vitamin niacin in CAC?

A. formation of NAD^+, it is the coenzyme for isocitrate dehydrogenase, α-ketoglutarate dehydrogenase complex, and malate dehydrogenase.

109. What is the role of vitamin thiamin in CAC?

A. Formation of TPP. It is coenzyme for pyruvate dehydrogenase and α-ketoglutarate dehydrogenase.

110. What is the role of vitamin pantothenic acid in CAC?

A. Pantothenic acid as a part of CoA. It is attached to carboxylic residues of acetyl CoA and succinyl CoA.

111. What are key enzymes of citric acid cycle?

A. Citrate synthase, isocitrate dehydrogenase and α-ketoglutarate dehydrogenase.

112. How CAC cycle is regulated?

A. Through availability of ATP, $NADH^+$, acetyl CoA, citrate, succinyl CoA, oxaloacetate, and long chain acyl CoA.

113. How can ATP and $NADH+H^+$ regulate citric acid cycle?

A. They inhibit all key enzymes of CAC.

114. What are mechanisms of regulation of citric acid cycle?

A. Citrate synthase is stimulated by acetyl CoA, oxaloacetate, ADP and NAD^+ and inhibited by long chain acyl CoA, citrate, succinyl CoA, ATP and $NADH+H^+$. Isocitrate dehydrogenase and α-Ketoglutarate dehydrogenase are stimulated by NAD^+ and ADP and inhibited by $NADH+H^+$ and ATP.

115. Why does citric acid cycle need oxygen to proceed?

A. Because in absence of oxygen respiratory chain is inhibited leading to increase $NADH+H^+/NAD$. $NADH+H^+$ will inhibit TCA cycle.

116. Pentose phosphate pathway use how much glucose?

A. About 10% of glucose molecules per day are entering in this pathway.

117. What is the purpose of pentose phosphate pathway?

A. It generates $NADPH+H^+$ and pentoses.

118. *What are the uses of pentoses in biological system?*

A. The pentose phosphates are necessary for synthesis of nucleic acids (DNA and RNA) and different nucleotidea.

119. *What is the use of $NADPH^+$ in biological systems?*

A. For reductive biosynthesis.

120. *What reductive biosynthesis pathways need $NADPH+H^+$?*

A. Fatty acid biosynthesis, synthesis of cholesterol, steroid hormones.

121. *What are the tissues in which pentose phosphate pathway is significant?*

A. Liver, adipose tissue, RBCs, adrenal cortex, ovary, testis, mammary glands, lens.

122. *Why these tissues?*

A. Because they need $NADPH+H^+$ for reductive biosynthesis.

123. *Apart from reductive synthesis, $NADPH+H^+$ is used for what purpose?*

A. It is necessary to keep the integrity of RBC membrane, for keeping glutathione in reduced state, for keeping transparency of lens, and it is necessary for superoxide production inside macrophages.

124. *Which is the key enzyme of pentose phosphate pathway?*

A. Glucose-6-phosphate dehydrogenase.

125. *What is the hormonal control over pentose phosphate pathway?*

A. Insulin stimulates the pathway by activating the key enzyme.

126. *What are other regulators for pentose phosphate pathway?*

A. The key enzyme is also stimulated by $NADP^+$ and inhibited by $NADPH+H^+$ and acetyl CoA.

127. *What are enzymes that generate $NADPH+H^+$?*

A. Glucose-6-phosphate dehydrogenase and 6-phosphogluconate dehydrogenase.

128. *What are sources of $NADPH+H^+$?*

A. Pentose phosphate pathway, action of malic enzyme on malate

and action of cytosolic isocitrate dehydrogenase on isocitrate.

129. *What about ATP generation? Is NADPH+H$^+$ is used for that?*

A. No. NADPH+H$^+$ is not used for ATP generation.

130. *What is the most common enzyme deficiency in man?*

A. Glucose-6-phosphate dehydrogenase deficiency.

131. *What is the disease resulting from glucose -6- phosphate dehydrogenase deficiency?*

A. Favism, where there are excessive hemolysis of RBCs → hemolytic anemia.

132. *Why deficiency of glucose-6-phosphate dehydrogenase enzyme causes anemia?*

A. Deficiency of glucose-6-P- dehydrogenase → ↓ NADPH,H$^+$ production → ↓ reduced glutathione → Accumulation of H_2O_2 → Hemolysis of RBCs?

133. *Why H_2O_2 causes hemolysis of RBCs?*

A. H_2O_2 causes peroxidation of fatty acids present in RBCs membrane and converts hemoglobin into met-hemoglobin. Peroxides and met-hemoglobin are toxic compounds that increase the red cell membrane fragility.

134. What is the mode of hereditary transmission of GPD deficiency?

A. It is transmitted as an x-linked recessive character.

135. Is there any advantage of the abnormal gene of glucose-6-phosphate dehydrogenase?

A. The geographical distribution of GPD deficiency correlates well with the malarial endemicity. The GPD deficiency offers resistance to malarial infection.

136. Acute hemolytic episode after administration of antimalarial drug is due to what?

A. Deficiency of glucose-6-phosphate dehydrogenase.

137. Transketolase activity is decreased in the deficiency of what?

A. Thiamine pyrophosphate (TPP).

138. *What is respiratory burst?*

A. Respiratory burst is the rapid consumption of molecular oxygen

due to its conversion into superoxide.

139. *What is the mechanism of respiratory burst?*

A. Phagocytic cells contain an enzyme called NADPH, H^+ oxidase enzyme. After phagocytosis of microorganisms has occurred, $NADPH^+,H^+$ oxidase converts oxygen (O_2) derived from surrounding tissues into superoxide ions (O_2^-). Super oxide then is converted into H_2O_2 and Hypochlorite (HOCl-) that Kill bacteria. This is called respiratory burst.

140. *What are the functions of uronic acid pathway?*

A. It is used for conjugation of bilirubin, steroids, synthesis of glucosaminoglycans.

141. *In lower animals as birds, uronic acid pathway is used for what purpose?*

A. For synthesis of ascorbic acid (vitamin C).

142. *What are fate of uridine diphosphate glucose (UDP-G)?*

A. Glycogen synthesis, glucuronic acid synthesis, amino sugar synthesis and lactose synthesis

143. *What is essential pentosuria?*

A. Excretion of pentose (L-xylulose) in urine due to the deficiency of xylitol dehydrogenase.

144. *What is its importance?*

A. It does not produce any harm. But it gives a positive reaction to Benedict's test, so it should be differentiated from diabetes mellitus.

145. *What is gluconeogenesis?*

A. Production of glucose from non-carbohydrate sources.

146. *What are those non-carbohydrate sources? (What are the substrates for gluconeogenesis?)*

A. Glucogenic amino acids, lactate, and glycerol.

147. *Is propionate a substrate for gluconeogenesis?*

A. Yes, but this occurs only in ruminants.

148. *Can fatty acids synthesize glucose, and why?*

A. No, because acetyl CoA cannot be converted into pyruvate (the

reaction pyruvate to acetyl CoA is a totally irreversible).

149. *Gluconeogenesis is taking place in which tissue?*

A. Liver (90%) and kidney (10%).

150. *Blood glucose level can be raised by glycogenolysis mainly by liver, why?*

A. Glucose-6-phosphatase is present only in liver.

151. *What is the significance of gluconeogenesis?*

A. Gluconeogenesis is necessary to maintain blood glucose level especially under conditions of starvation.

152. *What are functions of Gluconeogenesis?*

A. Supplies the body with glucose, which is essential source of energy during prolonged fasting and for tissues under anaerobic conditions, as nervous tissues, RBCs and skeletal muscles. Glucose is the precursor of milk sugar (lactose) in mammary gland. Gluconeogenesis clears the blood from the waste products of other tissues as lactate (produced by muscles and RBCs).

153. *What is pyruvate carboxylase?*

A. The enzyme catalyzing the reaction, pyruvate to oxaloacetate.

154. *What are the key gluconeogenic enzymes?*

A. Pyruvate carboxylase, phosphoenol pyruvate carboxykinase, Fructose-1,6- bisphosphatase and Glucose-6-phosphatase.

155. *Pyruvate carboxylase reaction (pyruvate to oxaloacetate) needs which coenzyme?*

A. Biotin and ATP.

156. *Malate shuttle is used for what purpose?*

A. Reactions of gluconeogenesis are taking place in cytosol. Hence, the oxaloacetate has to be transported from mitochondria to cytosol. This is achieved by the malate shuttle.

157. *What is the energy cost of conversion of 2 molecules of lactate into glucose (gluconeogenesis)?*

A. 6 ATP and 2 $NADH+H^+$

158. *What are the sources of these ATP molecules?*

A. (1) Two Pyruvate → Two oxaloacetate. (-2ATP), (2) Two Oxaloacetate → Two phosphoenolpyruvate (-2 ATP), (3) Two 3 Phosphoglycerate → Two 1,3 Bisphosphoglycerate (-2 ATP), (4) Two NADH+H$^+$: Two 1,3 bisphosphoglycerate → Two Glyceraldehyde-3-phosphate (2 NADH+H$^+$).

159. *What are mechanisms of regulation of gluconeogenesis?*

A. Hormonal regulation, ATP and acetyl CoA regulation.

160. *What are hormones stimulating gluconeogenesis?*

A. Glucocorticoids and glucagon.

161. *What are hormones inhibiting gluconeogenesis?*

A. Insulin.

162. *How can glucocorticoids regulate gluconeogenesis?*

A. They stimulate synthesis of all key enzymes of gluconeogenesis. Glucocorticoids stimulate also protein catabolism by tissues.

163. *What about insulin?*

A. Insulin inhibits gluconeogenesis. It acts as repressor (inhibitor) for synthesis of enzymes of gluconeogenesis.

164. *What are roles of ATP and acetyl CoA?*

A. ATP stimulates gluconeogenesis by stimulating fructose 1,6 bisphosphatase. Acetyl CoA stimulates pyruvate carboxylase.

165. *What are sources and fate of pyruvate?*

***Sources*: Glucose oxidation (glycolysis), lactate (by lactate dehydrogenase), malate (by malic enzyme), alanine (by transamination), serine (by non-oxidative deamination) and other amino acids (methionine, cysteine, threonine and glycine).**

***Fate:* Glucose formation (Gluconeogenesis), lactate formation (by lactate dehydrogenase), malate formation (by malic enzyme), alanine formation (by transamination), oxaloacetate formation (by pyruvate carboxylase).**

166. *What are functions of Glycogen?*

A. Liver glycogen maintains normal blood glucose concentration

especially during the early stage of fast (between meals). Muscle glycogen gives energy within the muscle itself especially during muscle contractions.

167. *What is glycogenolysis?*

A. Degradation of glycogen to glucose.

168. *Muscle glycogen will not serve as a precursor of blood glucose, why?*

A. Due to absence of glucose-6-phosphatase enzyme.

169. *What is the main enzyme for glycogenolysis?*

A. Glycogen phosphorylase.

170. *What will activate glycogen phosphorylase?*

A. Adrenalin, glucagon, cyclic AMP.

171. *What is the mechanism of action of adrenaline?*

A. Adrenaline increases cyclic AMP level, which activates glycogen phosphorylase.

172. *What is glycogenesis?*

A. Glycogen synthesis.

173. *What is the main enzyme for glycogenesis?*

A. Glycogen synthase.

174. *What will activate glycogen synthase?*

A. Insulin.

175. *What is the mechanism of action of insulin?*

A. Stimulation of phosphodiesterase enzyme ➔ Converts cAMP to AMP ➔ Stimulation of glycogenesis. Insulin also stimulates phosphatase enzymes ➔ Converts phosphorylated glycogen synthase into active dephosphorylated one ➔ Stimulation of glycogenesis.

176. *In the glycogen synthesis, which is the active glucose derivative?*

A. UDP-glucose.

177. *What are glycogen storage diseases?*

A. These are group of inherited disorders characterized by deposition

of abnormal type or quantity of glycogen in the tissues.

178. *What are causes of glycogen storage diseases?*

A. Deficiency of one of enzymes of glycogen metabolism.

179. *Give example of glycogen storage diseases?*

A. Type I or Von GierKe's disease.

180. *Which is the defective enzyme in Von Gierke's disease (glycogen storage disease type I)?*

A. Glucose-6-phosphatase.

181. *What is the main characteristic clinical feature of von Gierke's disease?*

A. Fasting hypoglycemia, which does not respond to adrenaline is very characteristic.

182. *What are manifestations of Von Gierke's disease?*

A. Fasting hypoglycemia, accumulation of large amounts of glycogen in liver, with subsequent disturbance of liver functions, enlargement of liver, ketosis, hyperlipidemia, and Hyperuricemia (↑ plasma uric acid).

183. *What are the functions of galactose?*

A. Galactose (UDP-galactose) enters in the structure of lactose (=milk sugar), glycolipids, glycoproteins and proteoglycans.

184. What is galactosemia?

A. It is increased blood galactose concentration due to inability to metabolize galactose.

185. What are the features of galactosemia?

A. Congenital cataract, mental retardation, neonatal hypoglycemia, hepatosplenomegaly, positive Benedict's test.

186. *What is the disease resulting from deficiency of galactose-1-phosphate uridyl transferase enzyme?*

A. Galactosemia.

187. *What are other causes of galactosemia?*

A. Deficiency of galactokinase and UDP-galactose epimerase.

188. What is the treatment policy in galactosemia?

A. Lactose free diet is given for first five years of life.

189. Why five years, why life-long treatment is not required?

A. By five years, the alternate pathway (galactose-1-phosphate pyrophosphorylase) becomes active.

190. How lactose is synthesized?

A. UDP glucose is epimerased to UDP galactose. Then the galactose unit is transferred from UDP-galactose to glucose. This synthesis of lactose in mammary gland is catalyzed by lactose synthase.

191. How lactose synthesis is regulated?

A. The lactose synthase has two subunits, a catalytic subunit which is a galactosyl transferase and a modifier subunit that is α-lactalbumin. The level of the modifier subunit is under the control of prolactin.

192. What is lactosuria?

A. It is observed in the urine of normal women during 3rd trimester of pregnancy and during lactation.

193. What is the clinical importance of lactosuria?

A. The condition is harmless. But it is important to distinguish lactosuria from glucosuria when gestational diabetes mellitus is suspected.

194. What are functions of fructose?

A. Energy production (15% of daily energy is derived from fructose), Fructose also is the major energy source for spermatozoa in the seminal vesicle.

195. Fructokinase catalyses which reaction?

A. Fructose to fructose-1-phosphate.

196. What is the disease resulting from deficiency of Aldolase B enzyme?

A. Hereditary fructose intolerance, where there is accumulation of fructose-1-phosphate.

197. What are manifestations of hereditary fructose intolerance?

A. Liver cell failure, renal failure and fasting hypoglycemia.

198. *What is the cause of fasting hypoglycemia?*

A. Due to inhibition of glycogen phosphorylase. This leads to inhibition of glycogenolysis and hypoglycemia.

199. *What is the disease resulting from deficiency of fructokinase enzyme?*

A. Essential fructosuria.

200. *Is essential fructosuria is harmful like hereditary fructose intolerance ?*

A. No, it is a benign metabolic defect due to deficiency of fructokinase. Urine gives positive Benedict's test, and so it should be differentiated from diabetes mellitus.

201. *Free fructose is seen in which body fluid?*

A. Seminal plasma.

202. *What is the clinical application of fructose estimation in semen?*

A. Fructose is secreted by seminal vesicles. A block in seminal vessels is indicated by the absence of fructose in semen.

203. *What is the level of fasting blood glucose in a normal person?*

A. 65-110 mg / dl.

204. *What is the level of blood glucose 2 hours after carbohydrate meal in a normal person?*

A. 65-140 mg / dl.

205. *What are sources of blood glucose?*

A. Dietary carbohydrate, liver glycogen, amino acids, and other metabolites (gluconeogenesis).

206. *How can regulation of blood glucose be achieved?*

A. By hepatic, hormonal, and renal regulations.

207. *What is the role of liver in regulation of blood glucose?*

A. After meal glucokinase and glycogen synthase are stimulated leading to glycogenesis. During fasting liver add blood glucose by glycogenolysis and gluconeogenesis.

208. *Which hormone is hypoglycemic?*

A. Insulin.

209. *Where is insulin synthesised?*

A. Beta cells of islets of Langerhans of pancreas.

210. *What are the anti-insulin (hyperglycemic) hormones?*

A. Glucagon, adrenaline, corticosteroids, and growth hormone.

211. *How is insulin secretion controlled?*

A. Glucose is the major stimulant of insulin secretion.

212. *What are the major actions of insulin?*

A. Insulin decreases blood sugar, it stimulates glucose uptake by cells, and glycolysis. It inhibits gluconeogenesis, glycogenolysis and lipolysis.

213. *What are the structural features of insulin?*

A. Insulin is a protein hormone with two polypeptide chains, the A chain with 21 amino acids and the B chain with 30 amino acids. Both chains are joined together by a pair of disulfide bonds. It has a total of 51 amino acids.

214. *What is the structural feature of insulin receptor?*

A. Insulin receptor has four subunits, two alpha and two beta subunits. The alpha units are located on the extracellular side, to which insulin binds. The beta subunits are towards cytoplasmic side. Beta subunit has tyrosine kinase activity.

215. *What are the major actions of glucagon?*

A. Glucagon increases blood sugar, it inhibits glycolysis. It stimulate glycogenolysis, gluconeogenesis and lipolysis.

216. *What are the pathways stimulated by insulin?*

A. Glycolysis, glycogen synthesis, pentose phosphate pathway, and lipogenesis.

217. *Name important enzymes that are stimulated by insulin.*

A. Phosphofructokinase, glycogen synthase, glucose-6-phosphate dehydrogenase, acetyl CoA carboxylase.

218. *What are the pathways inhibited by insulin?*

A. Glycogenolysis, gluconeogenesis, lipolysis, and ketogenesis.

219. What are the important enzymes inhibited by insulin?

A. Glucose-6-phosphatase, glycogen phosphorylase, hormone sensitive triacylglycerols lipase.

220. What does diabetes mean?

A. Diabetes means increased daily urine volume above normal concentration.

221. What are types of diabetes?

A. Diabetes mellitus, diabetes insipidus, bronze diabetes and diabetes innocence.

222. What is diabetes mellitus?

A. It is a state of chronic hyperglycemia, usually accompanied by glycosuria. It is caused by a relative or absolute deficiency of insulin hormone.

223. How diabetes mellitus is classified?

A. Type 1 and type 2.

224. What is the main diference between type I and Type II?

A. In type 1 there is absence of insulin, while in type 2 there is relative deficiency of insulin.

225. What are the characteristic features of type 1, diabetes mellitus?

A. Here circulating insulin level is deficient or absent. These patients are dependent on insulin injections. Onset is during childhood. Patients are undernourished. They are more prone to developing ketosis.

226. What about type 2 diabetes mellitus?

A. 80% of the patients belong to this type. There is a relative insulin deficiency. It is commonly seen in individuals above 40 years. These patients are less prone to developing ketosis.

227. What are the main symptoms of diabetes mellitus?

A. Polyuria, polydypsia, polyphagia and weight loss.

228. What is the reason for polyuria in diabetes mellitus?

A. When the blood glucose level exceeds the renal threshold, (180 mg/dl) glucose is excreted in urine. Due to osmotic effect, more water accompanies the glucose.

229. *What is the reason for polydypsia in diabetes mellitus?*

A. To compensate for this loss of water, thirst centre is activated, and more water is taken (polydypsia).

230. *What is the reason for weight loss in diabetes mellitus?*

A. Insulin deficiency causes excessive lipolysis. This would lead to loss of weight.

231. *What is the reason for polyphagia in diabetes mellitus?*

A. To compensate the loss of glucose and protein,patient takes more food.

232. *What are the criteria for diagnosing diabetes mellitus?*

A. Fasting plasma sugar is more than 126 mg/dl, or, if 2-hour post glucose load, value is more than 200 mg/dl.

233. *What is impaired glucose tolerance (IGT)?*

A. When fasting plasma glucose level is between 110 and 126 mg/dl and 2-hour post-glucose value is between 140 and 200 mg/dl.

234. *Can you diagnose diabetes mellitus based on random blood glucose estimation?*

A. Diabetes is diagnosed, if the random plasma sugar level is more than 200 mg/dl,

235. *When a standard oral glucose tolerance test was done, the blood glucose levels of the patient were found as: 0 min = 130 mg/dl and 120 min = 220 mg/dl. What will be your diagnosis?*

A. Diabetes mellitus.

236. *What is the major indication for doing an oral glucose tolerance test (OGTT)?*

A. Patient has symptoms suggestive of diabetes mellitus, but fasting blood sugar value is inconclusive (between 100 and 126 mg/dl).

237. *What are the WHO criteria for diagnosis of diabetes mellitus using oral glucose tolerance test?*

A. Fasting plasma glucose is greater than 126 mg/dl. At least, one of the intermediate (30, 60, 90 min) plasma specimens has plasma glucose greater than 200 mg/dl.

238. *How can renal threshold for a person be evaluated?*

A. By doing oral glucose tolerance test.

239. *What is the best method for glucose estimation?*

A. Glucose oxidase peroxidase (GOD-POD) method.

240. *What is normal renal threshold for glucose?*

A. 180 mg/100ml.

241. *What is glycosuria (glucosuria)?*

A. Presence of glucose in urine.

242. *What are types of glycosuria?*

A. Diabetic glycosuria (due to insulin deficiency), pregnancy glycosuria (in 20% of pregnancies), diabetes innocence duo to defect in tubular renal reabsorption of glucose and transient glycosuria.

243. *What is transient glucosuria?*

A. It may occur in some people due to emotional stress. Excessive secretion of catecholamines will produce hyperglycemia and resultant glucosuria. Once the stress is removed, the glucosuria disappears.

244. *What is renal glucosuria (diabetes innocence)?*

A. Here glucose is excreted in urine due to a lowering of renal threshold. The blood sugar levels are within normal limits.

245. *What are the reducing substances seen in urine?*

A. Glucose, fructose, lactose, galactose, pentoses, ascorbic acid, glucuronides.

246. *What is the test for reducing sugars in urine?*

A. Specific by using strips containing glucose oxidase or nonspecific as Benedict's test.

247. *What are the acute complications of diabetes mellitus?*

A. Ketoacidosis, hyperosmolar non-ketotic coma, lactic acidosis.

248. *What are the chronic complications of diabetes mellitus?*

A. Atherosclerosis, thrombosis, paralysis, gangrene, microangiopathy, nephrosclerosis, cataract, peripheral neuropathy.

249. *What is microalbuminuria?*

A. It is the presence of albumin in urine in concentration of 30 to 200 mg/day. It indicates the start of renal damage, atherosclerotic diseases and cardiovascular mortality. Albumin more than 200 mg/day indicates frank diabetic nephropathy.

250. *What is the difference between glycosylation and glycation?*

A. Enzymatic addition of any sugar to a protein is called "glycosylation" while non-enzymatic process is termed "glycation"

251. *What is the basis of glycation?*

A. When there is hyperglycemia, proteins in the body may undergo glycation. It is a non-enzymatic process. Glucose is added to the N-terminal amino group of proteins.

252. *What is the significance of glycated hemoglobin?*

A. The determination of glycated hemoglobin is not for diagnosis of diabetes mellitus, but for monitoring the response of treatment. It is unaffected by recent food intake or recent changes in blood sugar levels. An elevated glycohemoglobin indicates poor control of diabetes mellitus. The risk of retinopathy and renal complications are proportionately increased with elevated glycated hemoglobin.

253. *Why excessive intake of alcohol produces hypoglycemia?*

A. Because ethanol inhibits gluconeogenesis.

254. *What are types of coma in diabetes mellitus?*

A. ketotic coma, hyperglycemic-hyperosmolal coma, and lactic acidosis coma.

255. *May hypoglycemic coma occur in diabetic patients?*

A. Yes, if a diabetic patient is treated by *excessive amount of insulin*

256. *Neonatal hypoglycemia is seen in which conditions?*

A. Glycogen storage disease, type I, galactosemia, fructose intolerance.

257. *What are the high energy compounds formed through glucose oxidation that release energy upon hydrolysis?*

A. 1.3 Diphosphoglycerate (phosphate bond), phosphoenolpyruvate

(phosphate bond), acetyl CoA (sulfate bond), succinyl CoA (sulfate bond).

258. *What are congenital disorders in carbohydrate metabolism?*

A. Diabetes mellitus, lactose intolerance, pyruvate kinase deficiency, hexokinase deficiency, favism, essential pentosuria, galactosemia, essential fructosemia, hereditary fructose intolerance and Glycogen storage diseases e.g. Von Gierke's disease.

259. *In glycoproteins, carbohydrate residues are attached to which group of the polypeptide chain?*

A. Hydroxyl group of serine or threonine.

260. *What is fate of glucose-6-phosphate in the body?*

A. Glycolysis, pentose phosphate pathway, uronic acid pathway, glycogenesis and lactose synthesis.

261. *Compare between pentose phosphate pathway and glycolysis.*

	HMP Pathway	Glycolysis
Location	In certain cells	In all cells
Oxidation of glucose	Occurs in the first reaction	Phosphorylation occurs first then oxidation
Coenzymes	$NADP^+$	NAD^+
Energy	No energy production	2 or 8 ATP
CO_2	Produced	Not produced
Pentoses production	Produced	Not produced

262. *Compare between malate dehydrogenase and malic enzyme*

	Malate dehydrogenase	Malic enzyme
Substrate	Malate	Same
End product	Oxaloacetate	Pyruvate
Coenzyme	NAD^+	$NADP^+$
Location	Cytosol and mitochondria	Cytosol

263. *Compare between aerobic and anaerobic glycolysis.*

	Aerobic	Anerobic
End product	Pyurvate	Lactate
Energy	6 or 8 ATP	2 ATP
Regeneration of NAD^+	Through respiratory chain in mitochondria (Glycerophosphate and aspartate-malate shuttles)	Through lactate formation in cytosol (Pyruvate → Lactate)
Availability to TCA in mitochondria	Available, and 2 pyurvate can be oxidized to give 30 ATP	Not available as lactate is a cytosolic substrate

264. *Compare between lactose, lactase, lactate, lactate dehydrogenase and lactose synthase*

Substance	Function
Lactose	Milk sugar–disaccharide (glucose+ galactose)
Lactase	Intestinal enzyme. Digest lactose into glucose and galactose.
Lactate	End product of anaerobic glycolysis.
Lactate dehydrogenase	Enzyme that converts lactate into pyurvate and vice versa
Lactose synthase	Enzyme present in mammary gland. It catalyzes lactose formation.

265. *Compare between glycerophosphate shuttle and malate-aspartate shuttle.*

	Glycerophosphate shuttle	Malate- aspartate shuttle
Coenzymes	NAD^+ for cytoplasmic glycerophosphate dehydrogenase and FAD for mitochondrial glycerophosphate dehydrogenase.	NAD^+ for both cytoplasmic and mitochondrial malate dehydrogenase
Energy produced by respiratory chain	2 ATP	3 ATP
Organ location	Muscle and nerves	Liver and heart

266. *Compare between synthase and synthetase enzymes.*

	Synthase	Synthetase
Class	Transferase	Ligase
Utilization of ATP or GTP	Does not utilize high energy phosphate	Utilize high energy phosphate in the reaction
Example	Citrate synthase– Glycogen synthase	Glutamine synthetase

267. *Compare between liver glycogen and muscle glycogen.*

	LIVER GLYCOGEN	MUSCLE GLYCOGEN
Sources	• Blood glucose • Other hexoses e.g. fructose • Noncarbohydrate sources e.g. lactate	• Blood glucose only
Amount	120 g maximum	400 g maximum
Concentration	6 % of liver weight	1 % of muscle weight
Function	Supply all body cells with glucose	Private source of energy for muscles only.
End product	Glucose	Lactate
Effect of hormones		
• Insulin	Stimulates glycogenesis	Same
• Epinephrine	Stimulates glycogenolysis	Same
• Glucagon	Stimulates glycogenolysis	No effect

268. *Compare between types I and II of diabetes mellitus.*

	Type I (IDDM)	Type II (NIDDM)
Age of onset	Usually during childhood	Usually after age 35
Nutritional status	Undernourished	Obese
Prevalence	10-20%	80-90%
Genetic background	Moderate	Very strong
Defect or deficiency of insulin	No insulin due to destruction of β-cells	Decreased secreted insulin or insulin resistance

Ketosis	Common	Rare
Plasma insulin	Low or absent	Normal to high
Acute complications	Ketoacidosis	Hyperosmolar coma
Oral hypoglycemic drugs	No response	Responsive
Treatment with insulin	Always necessary	Usually not required

269. *Compare between glucokinase and hexokinase.*

	Glucokinase	Hexokinase
Site	Liver	All tissue cells
Affinity to glucose	Low (high Km)	High (low Km)
Substrate	Glucose only	Glucose,galactose and fructose
Effect of insulin	Induces synthesis of glucokinase	No effect
Effect of G-6-P	No effect	Inhibits hexokinase allosterically
Function	Acts in liver cells after meals. It removes glucose coming in portal circulation , converting it into G-6-P	It phosphorylates glucose inside the cells. This makes glucose concentration more in blood than inside the cells → Continuous supply of glucose for the tissues even in the presence of low blood glucose concentration.

MCQ, Matching, True and False and Completion

Select and encircle the most appropriate answer or completion:

1. *The action of α-amylase on dietary starch produces all the following compounds EXCEPT:*
   A. Maltose
   B. Lactose
   C. Amylodextrin
   D. Achrodextrin
   E. Isomaltose

2. *All the following about glycolysis is true EXCEPT:*
   A. NAD^+ is required as a coenzyme.
   B. There is a net energy production.
   C. Occurs in cytosol.
   D. CO_2 is produced.
   E. 2,3 Bisphosphoglycerate may be produced in RBCs.

3. *Which of the following comparison between hexokinase and glucokinase is FALSE:*
   A. Only hexokinase is inhibited by glucose-6-phosphate
   B. Only glucokinase is present in the brain
   C. Synthesis of glucokinase –and not hexokinase- is induced by insulin
   D. The Michaelis constant of hexokinase for glucose is much smaller than that of glucokinase
   E. Hexokinase is present in most tissues

4. *The activity of phosphofructokinase-1 can be decreased by all the following EXCEPT:*
   A. ATP
   B. Citrate
   C. Glucagon
   D. Fructose 2,6 bisphosphate

5. *An example of susbstrate level phosphorylation is the reaction catalyzed by:*
   A. Glucose-6-phosphate dehydrogenase
   B. Glycerldehyde-3-phosphate dehydrogenase
   C. Pyruvate kinase
   D. Pyruvate dehydrogenase complex
   E. Pyruvate carboxylase

6. *An enzyme not involved in glycolysis is:*
   A. Aldolase
   B. α-Glycerophosphate dehydrogenase
   C. Enolase
   D. Pyruvate kinase
   E. Phosphoglycerate mutase

7. *Fluoride is an inhibitor of which of the following enzymes of the pathway of glycolysis:*
   A. Hexokinase
   B. Aldolase
   C. Pyruvate kinase
   D. Enolase
   E. Phosphhexose isomerase

8. *The Cori cycle may be described as:*
   A. The interconversion between glycogen and glucose-1-phosphate
   B. The synthesis of alanine from pyruvae in skleal muscles and the synthesis of pyruvate from alanine in the liver
   C. The synthesis of urea in liver and degradation of of urea to carbon dioxide and ammonia by bacteria in gut
   D. The production of lactate from glucose in peripheral tissues with the resynthesis of glucose from lactate in liver
   E. None of the above

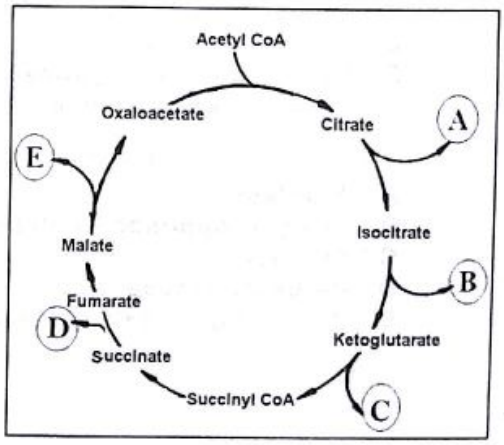
9. *Transport of glucose across the cell membrane is stimulated by insulin in:*
   A. Brain
   B. RBCs
   C. Liver
   D. Skeletal muscles

10. *Oxidative decarboxylation of pyruvate requires:*
   A. Thaimin pyrophosphate.
   B. $NADP^+$.
   C. Pyrodixal phosphate.
   D. Biotin.
   E. Cytochrome P 450.

11. *Pyruvate dehydrogenase complex enzyme is inhibited by the following EXCEPT:*
   A. Acetyl CoA
   B. ATP
   C. $NAD+H^+$
   D. Insulin
   E. Calcium ions

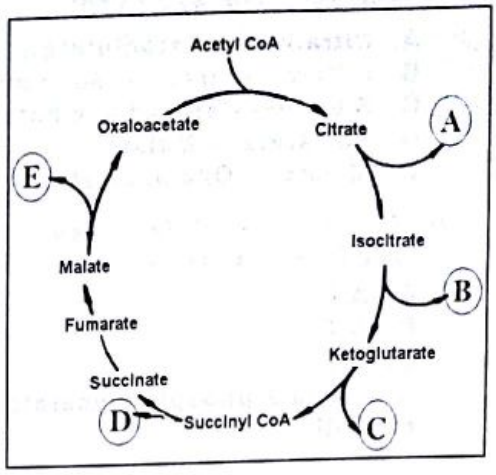
12. *Which of the following enzymes is NOT involved in the citric acid (krebs') cycle?*
   A. Fumarase
   B. Isocitrate dehydrogenase
   C. Succinate thiokinase
   D. Pyruvate dehydrogenase
   E. Aconitase

13. *All the lettered steps of citric acid cycle in the diagram shown beside donates a reduced coenzymes EXCEPT:*
   A. A
   B. B
   C. C
   D. D
   E. E



14. *Which step of citric acid cycle in the diagram shown below donates ATP by substrate level phosphorylation:*

A. A
B. B
C. C
D. D
E. E



15. *Which of the following statements about the citric acid cycle is <u>TRUE</u>:*

A. It contains no intermediates for gluconeogenesis
B. It contains intermediates for amino acids synthesis
C. It generates fewer molecules of ATP than glycolysis per mole of glucose consumed
D. It is an anaerobic process
E. It is a major anabolic pathway for pentose synthesis

16. *How many high energy compounds (ATP and GTP) are required to convert two molecules of lactate into glucose in mammalian liver*

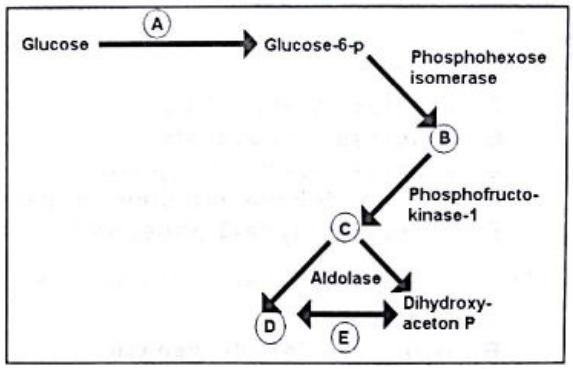
A. Two
B. Three
C. Four
D. Five
E. Six

17. *All the following substrates can be used to give glucose by gluconeogenesis <u>EXCEPT</u>:*

A. Aspartic acid
B. Glutamic acid
C. Succinyl CoA
D. Leucine
E. Glycerol

18. *In the figure shown beside, frucose 1,6 bisphosphate is located at point:*

A. A
B. B
C. C
D. D
E. E

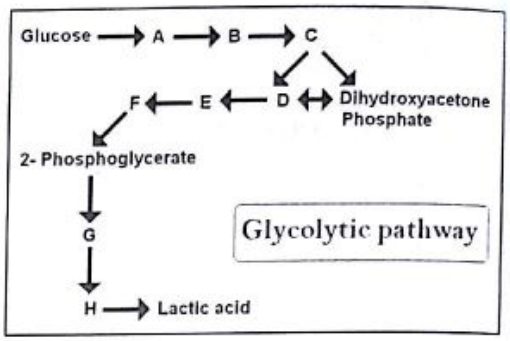


19. *In which of the following steps of tricarboxylic acid cycle, a high energy compound (ATP) is synthesized at the substrate level:*

A. Citrate → α-Ketoglutarate
B. α-Ketoglutarate → Succinyl CoA
C. Succinyl CoA → Succinate
D. Fumarate → Malate
E. Malate → Oxaloacetate

20. *In the pathway beside, ATP is produced between:*

A. A-B
B. B-C
C. C-D
D. F and 2-phosphoglycerate
E. G-H



21. *Which of the following metabolic pathway DOES NOT occur in mitochondria?*

A. Citric acid cycle
B. Electron transport chain
C. Glycolysis
D. Oxidative decarboxylation of pyruvate
E. Oxidative decarboxylation of α-ketoglutarate

22. *All the following compounds contain a high energy phosphate bond EXCEPT:*

A. ADP
B. Creatine phosphate
C. Glucose-6-phosphate
D. Phosphoenol pyruvate
E. 1,3 Bisphosphoglycerate

23. *The net number of ATP molecules formed per molecule of glucose in aerobic oxidation in hepatic cell is:*

A. 2
B. 8
C. 18
D. 38
E. 54

24. *The fructokinase reaction produces which of the following intermediates?*

A. Fructose-1-phosphate
B. Fructose-6-phosphate
C. Fructose-1,6 bisphosphate
D. Glyceraldehyde and dihydroxyacetone phosphate
E. Glyceraldehyde-3-phosphate and dihydroxyacetone phosphate

25. *All the following enzymes involved in citric acid cycle EXCEPT:*

A. Fumarase
B. Isocitrate dehydrogenase
C. Succinate thiokinase
D. Pyruvate dehydrogenase
E. Aconitase

26. *All of the following are intermediates of tricarboxylic acid cycle EXCEPT:*
   A. Citrate
   B. Malate
   C. Propoinyl CoA
   D. α-Ketoglutarate
   E. Oxaloacetate

27. *Which of the following is NOT enzyme of gluconeogenesis:*
   A. Glucose-6-phosphatase
   B. Fructose 1,6 bisphosphatase
   C. Phosphenol pyruvate carboxykinase
   D. Pyruvate kinase
   E. Pyruvate carboxylase

28. *cAMP activates:*
   A. Glycogen synthase
   B. Hexokinase
   C. Protein kinase
   D. Adenylate cyclase
   E. Phosphofructokinase-1

29. *All the following are true about glucuronic acid EXCEPT:*
   A. It is conjugated to some compounds making them more soluble before excretion
   B. It forms bilirubin glucuronides
   C. It enters in the synthesis of glycosaminoglycans
   D. It is a precursor of L-ascorbic acid in man
   E. End product is L-xylulose which then join the pentose phosphate pathway.

30. *Galactosemia results from deficiency of :*
   A. Glucokinase
   B. Fructose 1,6 bisphosphatase
   C. UDP-glucuronyl transferase
   D. Galactose-1-phosphate uridyl transferase
   E. Galactose reductase

31. *Gluconeogenesis enzymes include all the following EXCEPT:*
   A. Frucose 1,6 bisphosphatase
   B. Glucose-6-phosphatase
   C. Phosphoenolpyruvate carboxykinase
   D. Phosphoglucomutase
   E. Pyruvate carboxylase

32. *A substance that is NOT an intermediate in the formation of glucuronic acid from glucose is:*
   A. UDP-galactose
   B. UDP-glucose
   C. Glucose-6-phosphate
   D. UDP-glucuronic acid
   E. Glucose-1-phosphate

33. *Essential pentosuria is characterized by:*

A. Excretion of arabinose.
B. Excretion of ribitol
C. Excretion of L-xylulose.
D. Fructosuria.
E. Galactosuria.

34. *Dehydrogenases of pentose phosphate pathway require:*

A. NAD^+
B. $NADP^+$
C. FAD
D. FMN
E. TPP

35. *The major process responsible for maintaining blood glucose 40 hours after last meal is:*

A. Glycolysis
B. Uronic acid pathway
C. Glycogenolysis
D. The pentose phosphate pathway.
E. Gluconeogenesis

36. *The major process responsible for maintaining blood glucose 4 hours after last meal is:*

A. Glycolysis
B. Uronic acid pathway
C. Glycogenolysis
D. The pentose phosphate pathway.
E. Gluconeogenesis.

37. *Fructose and galactose enter the liver and are phosphorylated at carbon:*

A. 1
B. 2
C. 3
D. 5
E. 6

38. *Each of the following enzymes is required for the conversion of glycerol to glucose EXCEPT:*

A. Glucose-6-phosphatase
B. Glycerol-3-phosphate dehydrogenase
C. Phosphoenol pyruvate carboxykinase
D. Fructose 1,6 bisphosphatase
E. Phosphohexose isomerase

39. *Which of the following enzymes of glycolysis is utilized in gluconeogenesis:*

A. Glucokinase
B. Phosphofructokinase
C. Pyruvate kinase
D. Aldolase

40. *Galactosemia is caused by:*
   A. The high content of lactose in artificial feeding formulae for babies.
   B. Absorption of none digested lactose through intestinal mucosa.
   C. Excessive conversion of gluose-1-phosphate to galactose-1-phosphate.
   D. Deficiency of galactose-1-phosphate uridyl transferase.
   E. Deficiency of UDP glucuronyl transferase.

41. *In gluconeogenesis each of the following is required for the conversion of pyruvate to phosphoenol pyruvate EXCEPT:*
   A. Phosphoenol pyruvate carboxykinase.
   B. GTP.
   C. Pyruvate carboxylase.
   D. Pyruvate kinase.
   E. CO_2

42. *How many net molecules of ATP are generated when one molecule of glucose is oxidized to lactate:*
   A. 0
   B. 1
   C. 2
   D. 12
   E. 38

43. *In which compartment does glycolysis occur?*
   A. The mitochondria
   B. The nucleus
   C. Cytosol
   D. The rough endoplasmic reticulum
   E. The smooth endoplasmic reticulum

44. *The following share in regulation of glycogen metabolism EXCEPT:*
   A. cAMP
   B. Protein kinase
   C. Phosphodiestrase
   D. Adenylate cyclase
   E. Acetyl CoA

45. *All the following about glycogenolysis are true EXCEPT:*
   A. It is the breakdown of glycogen
   B. The end product is glucose in both liver and muscles
   C. It is stimulated by glucagon
   D. Von-Gierk's disease results from deficiency of glucose-6-phosphatase
   E. Phosphrylase acts on more than 4 glucosyl units branch

46. *All the following about the pentose phosphate pathway are true EXCEPT:*
   A. There is a net energy production of 8 ATP molecules.
   B. $NADPH+H^+$ is produced that is essential for lipids metabolism.
   C. Pentoses are produced.
   D. Deficiency of glucose-6-phosphate dehydrogenase leads to favism.
   E. Its intracellular location is cytosol.

47. *In glycolysis, NAD^+ can be regenerated in the cytosol if $NADH+H^+$ reacts with following EXCEPT:*
   A. Dihydroxyacetone phosphate
   B. Oxaloacetate
   C. Pyruvate
   D. Lactate

48. *In glycolysis, one of enzymes that catalyzes the synthesis of ATP is:*
   A. Hexokinase
   B. Glucokinase
   C. Phosphofruktokinase-1
   D. Glyceraldehyde-3-phosphate dehydrogenase
   E. Phosphoglycerate kinase

49. *One of irreversible reactions of glycolysis is that catalyzed by:*
   A. Phosphohexose isomerase
   B. Phosphofructokinase-1
   C. Aldolase
   D. Glyceraldehyde –3-phosphate dehydrogenase
   E. Phosphoglycerate mutase

50. *Glucokinase:*
   A. Is found in muscles
   B. Has low affinity (high Km) to glucose
   C. Allosterically inhibited by glucose-6-phosphate
   D. Catalyzes a reversible reaction
   E. Its synthesis is induced by glucagon

51. *Which statement concerning the well fed state is CORRECT:*
   A. Most enzyme is active in a phosphorylated form
   B. Hepatic fructose 2,6 bisphosphate is elevated
   C. Synthesis of glucokinase is repressed
   D. Synthesis of pyruvate dehydrogenase is repressed
   E. Glycogenolysis is stimulated

52. *Actions of fructose 2,6 bisphosphate include:*
   A. Inhibition of pyruvate dehydrogenase
   B. Stimulation of phosphofructokinase I
   C. Stimulation of phosphofructokinase II
   D. Stimulation of glucokinase
   E. Inhibition of pyruvate kinase

53. *Complete oxidation of one molecule of glucose under anaerobic glycolysis gives:*
   A. 2ATP
   B. 6 ATP
   C. 8 ATP
   D. 30 ATP
   E. 38 ATP

54. *In hepatic cells, the first step of glycolysis is catalyzed by:*
   A. Glucokinase
   B. Fructokinase
   C. Galactokinase
   D. Pyruvate kinase
   E. Glucose-6-phosphate dehydrogenase

55. *Von Gierke's disease is caused by defective:*
- A. Amylo α 1,6 glucosidase.
- B. Branching enzyme
- C. α 1,4 glucosidase.
- D. Phosphorylase.
- E. Glucose-6-phosphatase.

56. *Hereditary fructose intolerance is due to:*
- A. Deficiency of aldolase A
- B. Deficiency of aldolase B
- C. Deficiency of fructokinase
- D. Deficiency of fructose reductase
- E. Deficiency of hexokinase

57. *Anaerobic glucose oxidation produces:*
- A. One molecule of pyruvic acid
- B. Two molecules of pyruvic acid
- C. One molecule of lactic acid
- D. Two molecules of lactic acid
- E. Three molecules of lactate

58. *Degradation of liver glycogen gives:*
- A. Lactic acid
- B. Pyruvic acid
- C. Glucose
- D. Acetyl CoA
- E. Glucuronic acid

59. *Oxidation of acetyl CoA in Krebs' cycle produces:*
- A. ATP
- B. 12 ATP
- C. 15 ATP
- D. 24 ATP
- E. 38 ATP

60. *Pentose phosphate pathway produces:*
- A. ATP
- B. CO_2
- C. $NADPH+H^+$
- D. $NADH+H^+$
- E. $FADH_2$

61. *Anemia associated with G-6-phosphate dehydrogenase deficiency is characterized by:*
- A. Increased ratio of reduced to oxidized glutathione
- B. Increased ratio of $NADPH^+$ to $NADP^+$
- C. Peroxidation of RBCs cell membrane fatty acids
- D. Prevented by diets as fava been.
- E. All of the above

62. *Glucokinase is an example of:*
- A. Oxidoreductases
- B. Transferases
- C. Hydrolases
- D. Isomerases
- E. Ligases

63. *Oxidation of citrate to succinate results in the production of how many moles of high energy phosphate bonds per mole of citrate oxidized?*
   A. 4
   B. 5
   C. 6
   D. 7
   E. 8

64. *Is (are) used to maintain blood glucose levels during fasting:*
   A. Muscle glycogen
   B. Liver glycogen
   C. Both of them.
   D. Neither of them

65. *Kinases require:*
   A. Mn^{++}
   B. Mg^{++}
   C. Inorganic phosphate
   D. EDTA
   E. Epinephrin

66. *All the following enzymes catalyze a substrate level phosphorylation reactions EXCEPT:*
   A. Glucokinase
   B. Succinate thiokinase
   C. Pyruvate kinase
   D. Phosphoglycerate kinase

67. *Which one of the following statements is characteristic of gluconeogenesis?*
   A. It does not utilize energy in the form of ATP or GTP.
   B. It maintains blood glucose during the long term fast.
   C. It involves the function fructose 1,6 bisphosphatase.
   D. It uses carbon skeleton provided by degradation of any amino acid.
   E. It is stimulated by insulin hormone.

68. *Epinephrine has which of the following effects on glycogen metabolism in the liver?*
   A. The net synthesis of glycogen is increased.
   B. Glycogen phosphorylase is inactivated whereas glycogen synthase is activated.
   C. Both glycogen phosphorylase and glycogen synthase are activated.
   D. Glycogen phosphorylase is activated whereas glycogen synthase is inactivated.
   E. Has no effect on liver glycogen.

69. *Which of the following is NOT a substrate for gluconeogenesis:*
   A. Alalnine.
   B. Stearate.
   C. A Ketoglutarate.
   D. Glutamate.
   E. Pyruvate.

70. *One of key enzymes in gluconeogenesis is:*
A. Aldolase
B. Enolase
C. Pyruvate kinase
D. Pyruvate carboxylase
E. Glucokinase

71. *Which of the following substrates CANNOT be converted into glucose:*
A. Lactate
B. Glycerol
C. Acetyl CoA
D. Aspartate
E. Oxaloacetate

72. *A conversion of glucose-6-phosphate into lactate in glycolysis is a accompanied by a net gain of:*
A. One mole of ATP
B. Two moles of ATP
C. Three moles of ATP
D. 6 moles of ATP
E. 8 moles of ATP

73. *The enzyme which is lacking in muscles but present in normal liver is:*
A. Glycogen phosphorylase
B. Phosphglucomutase
C. Glucokinase
D. Glucose-6-phosphatase
E. Creatine kinase Isoenzyme MM

74. *In muscle glycogenolysis, what is the net ATP given in converting one glucosyl residue in glycogen to two molecules of lactate?*
A. One
B. Two
C. Three
D. Four
E. Five
F. Six

75. *In hepatic cells, the ratio of the number of ATP molecules formed per mole of glucose under aerobic conditions to the number formed per mole of glucose under anaerobic conditions in red cells is:*
A. 2:1
B. 9:1
C. 15:1
D. 19:1
E. 38:1

76. *Enzymes which are active in the dephosphorylated form DO NOT include:*
A. Glycogen synthase
B. Glycogen phosphorylase
C. Pyruvate dehydrogenase
D. Acetyl CoA carboxylase
E. HMG CoA reductase

77. *The term "diabetes" means:*
A. Increased blood glucose
B. Decreased blood glucose
C. Increased daily urine glucose
D. Increased daily urine volume
E. Deficiency of insulin

78. *Diabetes mellitus is due to the deficiency of the action of which hormones:*
A. Glucagon
B. Glucocorticoids
C. Nor-epinephrine
D. Insulin
E. Thyroxin

79. *In a symptomatic patient, diabetes mellitus can be diagnosed by estimation of:*
A. Glycated hemoglobin
B. Detection of glucose in urine
C. Plasma level of C-peptide
D. Fasting and postprandial blood glucose
E. Microalbuminuria

80. *Glycosuria may be caused by the following EXCEPT:*
A. Diabetes mellitus.
B. Diabetes insipidus
C. Glycosuria of pregnancy.
D. Low renal threshold for renal tubular reabsorption of glucose.

81. *Which of the following blood glucose concentration is diagnostic of diabetes mellitus:*
A. Fasting plasma glucose = 160 mg/dl
B. Fasting plasma glucose = 115 mg/dl
C. Post prandial plasma glucose = 135 mg/dl
D. Postprandial plasma glucose = 160 mg/dl
E. Random plasma glucose = 80 mg/dl

82. *Normal fasting blood glucose concentration is:*
A. Less than 45 mgldl
B. 65-110 mg/dl
C. 110-126 mg/dl
D. 126-200 mg/dl
E. Above 200 mgldl

83. *Hypoglycemia is caused by:*
A. Insulinoma
B. Insulin resistant
C. Insulin deficiency
D. Diabetes insipidus
E. Diabetes mellitus

84. *Blood glucose level increases as a result of:*
A. Diabetes insipidus
B. Diabetes mellitus
C. Diabetes innocence
D. Glycosuria
E. Galactosemia

85. *Early renal impairment in diabetes mellitus can be expected by estimation of:*

A. Glycated hemoglobin
B. Detection of glucose in urine
C. Plasma level of C-peptide
D. Fasting and postprandial blood glucose
E. Microalbuminuria

86. *Long term diabetic control (2 months) can be checked by estimation of:*

A. Glycated hemoglobin
B. Fructoseamine
C. Detection of glucose in urine
D. Fasting and postprandial blood glucose
E. Microalbuminuria

87. *Diabetic control in pregnancy can be diagnosed by estimation of:*

A. Glycated hemoglobin
B. Fructoseamine
C. Detection of glucose in urine
D. Fasting and postprandial blood glucose
E. Microalbuminuria

88. *All the following statements about blood glucose levels in humans are true EXCEPT:*

A. Normal fasting values are 65-110 mg/dl
B. Average renal threshold for glucose are 180 mg/dl
C. After ingestion of large amounts of sugar, blood glucose levels may reach 200 mg/dl in normal individuals
D. 2 Hours postprandial blood sugar levels decrease from peak values more rapidly in normal individuals than in diabetics
E. Normal 2 hours postprandial values may exceed fasting diabetic values

In the following questions indicate with clear (T) the true statements, and with clear (F) the false statements:

Regulatory mechanism of glycolysis include:

89. The activation of phosphofructokinase-1 by ATP
90. The inhibition of glucokinase by glucose-6-phosphate
91. The inactivation of pyruvate kinase when glucagon levels are elevated
92. The activation of phosphofructokinase-1-by fructose 2,6 bisphosphate
93. The activation of glucokinase by insulin

The conversion of galactose to glucose requires:

94. A pyrophosphorylase
95. An epimerase
96. UDP-glucuronyltransferase
97. Galactose-1-phosphate uridyl transferase
98. A galactokinase

In In tricarboxylic acid cycle (TCA) cycle:

99. 12 ATP are produced through respiratory chain phosphorylation.
100. Succinyl CoA may be used in heme synthesis
101. $NADH+H^+$ and ATP stimulate TCA cycle.
102. CO_2 produced is used in carboxylation reactions.
103. There are intermediates which can be converted into amino acids

Pyruvate dehydrogenase complex:

104. Is an enzyme that catalyzes the conversion of pyruvate into lactate
105. Is inhibited by ATP, $NADH+H^+$, acetyl CoA and calcium ions
106. Needs 5 coenzymes, all are vitamin B complex derivatives
107. Is an enzyme catalyzes a reaction that produces 2 ATP
108. Is present in an active phosphorylated and inactive dephosphorylated forms

Hexokinase is an enzyme that:

109. Present in liver cells only.
110. Has high affinity (low Km) for glucose.
111. Allosterically inhibited by glucose-6-phosphate.
112. Its substrate is only glucose.
113. Maintains concentration gradient between blood and inside cells.

Lactate is the end product of anaerobic glycolysis:

114. May be converted into glucose again through lactic acid (Cori) cycle.
115. May be converted into pyruvate if oxygen gets available.
116. May be converted into pyruvate by reaction catalyzed by pyruvate dehydrogenase.
117. May cause lactic acidosis if its level is decreased.
118. Its production is essential for regeneration of oxidized NAD^+.

Pentose phosphate pathway:

119. NAD^+ is used as a coenzyme
120. Is an alternative pathway for glucose oxidation that produces energy.
121. Its metabolic error is essential pentosuria.
122. Glucose -6- phosphate dehydrogenase enzymes catalyze its committed steps.
123. Produces pentoses essential for synthesis of nucleic acids and different nucleotides.

Phosphofructokinase-2:

124. An enzyme that catalyzes the formation of fructose 2,6 bisphosphate
125. An enzyme that catalyzes an irreversible reaction.
126. Its product has a role in regulation of both glycolysis and gluconeogenesis.
127. An enzyme that is present mainly in liver.
128. May act as a phosphatase enzyme.

Glycogen:

129. Is mainly present in adipose tissue.
130. Maintains blood glucose levels between meals.
131. Is depleted from liver after 6 hours.
132. Is homopolysaccharide formed of glucose polymer.
133. Its synthesis is stimulation by epinephrine and nor epinephrine

Gluconeogenesis:

134. Occurs in brain and muscles.
135. Is essential for production of glucose after short term fast.
136. Is stimulated by insulin and inhibited by glucocorticoids.
137. Is a pathway that is exactly the reverse of glycolysis.
138. Is an anabolic process that utilizes 6 high energy phosphate bonds for conversion of 2 pyruvate into glucose.

Type I diabetes:

139. Occurs usually early in life.
140. The patient is usually obese.
141. Is due to destruction of β cells of islets of Langerhans.
142. Rarely proceeds to ketosis.
143. Treatment with insulin is always necessary.

Type II diabetes:

144. Occurs frequently after the age of 35 years.
145. Plasma insulin is absent.
146. The patient is usually thin.
147. Genetic background is strong.
148. Treatment with insulin is always necessary.

The following are causes of hypoglycemia:

149. Insulinoma
150. Diabetes mellitus
151. Severe liver disease
152. Hyper-pituitarism
153. Leucine hypersensitivity in infants.

High energy bonds are found in:

154. Glyceraldehyde-3-phosphate
155. Phosphoenol pyruvate
156. ATP
157. Creatine phosphate
158. Succinyl CoA

Galactose-1-phosphate uridyl transferase:

159. Essential for galactose metabolism.
160. Its deficiency is usually associated with fructosemia.
161. Its substrate is galactose.
162. Its deficiency leads to mental retardation, liver cell failure and cataract

Uronic acid pathway:

163. Needs both NAD^+ and $NADP^+$
164. Is essentially for production of glucuronic acid.
165. Its metabolic error is essential pentosuria.
166. In human it produces vitamin C.
167. Occurs in cytosol of liver cells

Which of the following statements about the structure of glycogen is TRUE:

168. There are α 1,4 glycosidic linkage
169. There are α 1,6 glycosidic linkage
170. All the monosaccharides in glycogen are α-D-glucose
171. Glycogen is an unbranched molecules

The reaction catalyzed by α-Ketoglutarate dehydrogenase in citric acid cycle requires:

172. NAD^+
173. $NADP^+$
174. FAD
175. CoA
176. TPP

The following compounds are substrate for gluconeogenesis:

177. Oleic acid
178. Serine
179. Leucine
180. Glycerol
181. Lactate

The pentose phosphate pathway includes the following enzymes:

182. Glucose-6-phosphate dehydrogenase
183. Glucose-6-phosphatase
184. Transketolase
185. Transaldolase
186. Aldolase

Oxidative decarboxylation of pyruvate requires:

187. NAD^+
188. $NADP^+$
189. FAD
190. Folic acid
191. Cobalamine

The citric acid cycle is inhibited by:

192. Arsenate
193. Malonate
194. Flouroacetate
195. Iodoacetate
196. Flouride ions

The following are inhibitors of phosphofructokinase-1 enzyme:

197. Citrate
198. Insulin
199. ATP
200. Cyclic AMP
201. Fructose 2,6 bisphosphate

After digestion of a piece of cake that contains primarily flour, milk and sucrose, the magor carbohydrate product(s) enter portal circulations is (are):

202. Glucose
203. Fructose
204. Maltose
205. Galactose
206. Ribose

The cytosolic NADH+H$^+$ is transported into mitochondria for oxidation in respiratory chain through:

207. $NADH+H^+$ is transported across the mitochondrial membranes directly.
208. glycerophosphate shuttle
209. Carnitine shuttle
210. Malate aspartate shuttle
211. Creatine shuttle

The following statements are concerned with regulatory effects of citrate:

212. It inhibits phosphofructokinase-1
213. It activates acetyl CoA carboxylase
214. It activates enolase
215. It activates pyruvate kinase
216. It inhibits citrate synthase.

High energy phosphate bonds are found in:

217. Phosphenol pyruvate
218. ATP
219. Creatine phosphate
220. AMP
221. Glyceraldehyde -3- phosphate

Matching: For each set of numbered questions, choose the ONE BEST answer from the list of lettered options below it. An answer may be used once or more times, or not at all.

222. *Phosphoenol pyruvate carboxykinase*
223. *UDP-glucuronyl transferase*
224. *Glucose-6-phosphate dehydrogenase*
225. *Pyruvate dehydrogenase complex*

A. Bilirubin catabolism
B. Gluconeogenesis
C. Oxidative decarboxylation of pyruvate
D. Pentose phosphate pathway

226. *Complete oxidation of one molecule of glucose in liver cell*
227. *Oxidation of one molecule of glucose in RBCs*
228. *Complete oxidation of one molecule of pyruvate*
229. *Complete oxidation of one molecule of Acetyl CoA*

A. 2 ATP.
B. 12 ATP
C. 15 ATP
D. 38 ATP.

230. *Lactose*
231. *Lactate*
232. *Lactate dehydrogenase*
233. *Lactase*

A. One of intestinal disaccharidases.
B. An enzyme that can be used in diagnosis of myocardial infarction.
C. Milk sugar.
D. End product of anaerobic glycolysis

234. *Glycogen phosphorylase*
235. *Glucokinase*
236. *Glucose-6-phosphatase*
237. *Phosphofructokinase-2*

A. Lacking in Von Gierk's disease
B. Catalyzes phosphorolysis of glycogen
C. Bifunctional enzyme
D. Catalyzes first step of glycolysis

238. *Deficiency of aldolase B*
239. *Deficiency of glucose-6-phosphate dehydrogenase*
240. *Deficiency of galactose-1-phosphate uridyl transferase*
241. *Deficiency of lactase*

A. Favism
B. Hereditary fructose intolerance
C. Lactose intolerance
D. Galactosemia

242. *Glycogenesis*
243. *Glycogenolysis*
244. *Glycolysis*
245. *Gluconeogenesis*

A. Synthesis of glucose from non-carbohydrate sources
B. Breakdown of glycogen into glucose
C. Synthesis of glycogen
D. Breakdown of glucose into lactate

246. *Glucose-6 phosphate → Glucose*
247. *Glucose 6-phosphate → 6 Phosphogluconolactone*
248. *Glucose-6-phosphate → Fructose-6 phosphate*
249. *Glucose-6-phosphate → 6 Phosphoglcuronic acid*

A. Glycolysis
B. Uronic acid pathway
C. Pentose phosphate pathway
D. Gluconeogenesis

250. *Pyruvate → Acetyl CoA*
251. *Pyruvate → Oxaloacetate*
252. *Phosphoenol pyruvate → Pyruvate*
253. *Pyruvate ⇆ Lactate*
254. *Pyruvate ⇆ Alanine*

A. Pyruvate kinase
B. Pyruvate dehydrogenase complex
C. Pyruvate carboxylase
D. Lactate dehydrogenase
E. ALT

255. *Pyruvate + CO_2 + Biotin + ATP → Oxaloacetate + ADP+ Pi*
256. *Galactose-1-phosphate + UDP-glucose → UDP-galactose + Glucose-1-phosphate*
257. *Acetyl CoA + Oxaloacetate + H_2O → Citrate + CoASH*
258. *Pyruvate + TPP+ Lipoic acid + CoASH + FAD + NAD^+ → Acetyl CoA + 3 ATP*
259. *Fructose-6-phosphate + ATP → Fructose 2,6 bisphosphate + ADP*

A. Pyruvate dehydrogenase complex
B. Pyruvate carboxylase
C. Phosphofructokinase-2
D. Citrate synthase
E. Galactose—1-phosphate uridyl transferase

260. *Pyruvate + $NADH+H^+$ ↔ Lactate + NAD^+*
261. *Glucose-6-phosphate + $NADP^+$ → 6 Phosphogluconolactone + $NADPH+H^+$*
262. *O_2 + $NADPH+H^+$ → O_2^- (superoxide) + $NADP^+$*
263. *UDP-glucose + Glycogen primer → α 1-4 glucosyl units + UDP*
264. *Glucose-6-phosphate → Glucose + Pi (inorganic phosphate)*

A. $NADPH+H^+$ oxidase.
B. Glucose-6-phosphatase
C. Glucose-6-phosphate dehydrogenase
D. Lactate dehydrogenase.
E. Glycogen synthase

265. *Glucose-6-phosphate dehydrogenase*
266. *Aldolase*
267. *Isocitrate dehydrogenase*
268. *Glycogen synthase*

A. Produces compounds that contains 3 carbons.
B. Produces α 1,4 glucosyl units
C. Produces $NADPH+H^+$
D. Produces $NADH+H^+$

269. Carboxylation of propionate
270. Transamination of amino acids
271. Transketolation
272. Activation of palmitate
273. Oxidation of lactate

A. Thiamin
B. Pantothenic acid
C. Riboflavin
D. Nicotinic acid
E. Folic acid
F. Pyridoxine
G. Biotin

274. Deficiency of pyruvate kinase.
275. Failure of conversion of L-xylose into D xylose
276. Insulinoma
277. Increased blood lactate above normal concentration

A. Hypoglycemia
B. Hemolytic anemia
C. Acidosis
D. Essential pentosuria

Match the numbered reaction and the lettered inhibitor:

278. Glucose → Glucose -6- phosphate
279. Fructose -6- phosphate → Fructose -1,6- bisphosphate
280. Fructose -1,6- bisphosphate → Fructose -6- phosphate
281. Phosphoenolpyruvate → Pyruvate
282. 2 Phosphoglycerate → Phosphoenol pyruvate

A. Citrate
B. Fructose -1,6- bisphosphate
C. Insulin
D. Glucose -6- phosphate
E. Fluoride salt

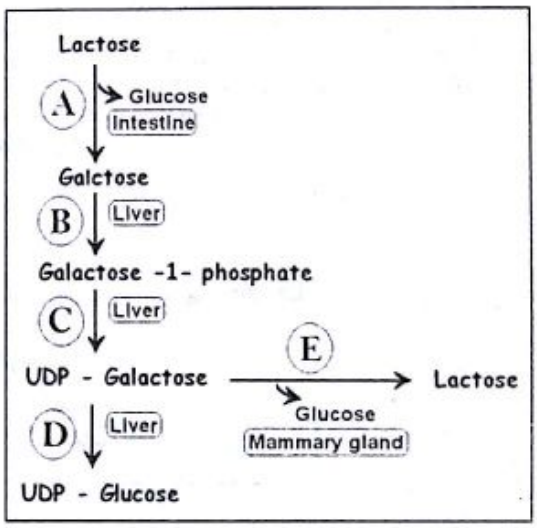
Match the numbered inhibitor and the lettered enzyme:

283. Flouride
284. Flouroacetate
285. Flourooxaloacetate
286. Malonate
287. Aresenate

A. Succinate dehydrogenase
B. α-Ketoglutarate dehydrogenase
C. Enolase
D. Citrate synthase
E. Malate dehydrogenase

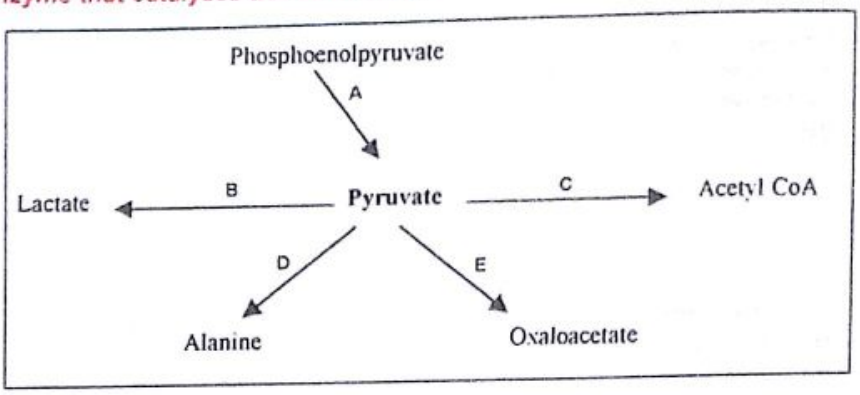
For each numbered enzyme, select the lettered reaction that is catalyzed by that enzyme in the diagram below:

288. Lactose synthase
289. Lactase
290. Galctokinase
291. UDP-Galactose epimerase
292. Galactose -1- phosphate uridyl transferase



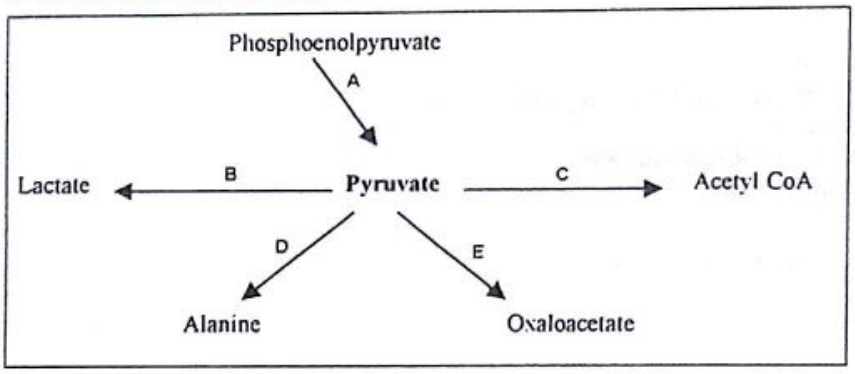
For each num bered enzyme, select the lettered reaction in the figure below that is catalyzed by that enzyme in the diagram below:

293. *An enzyme for gluconeogenesis*
294. *An enzyme that needs 5 coenzymes*
295. *An enzyme that has 5 isoenzymes*
296. *An enzyme that generates a net one ATP molecule*
297. *An enzyme that catalyzes transamination*



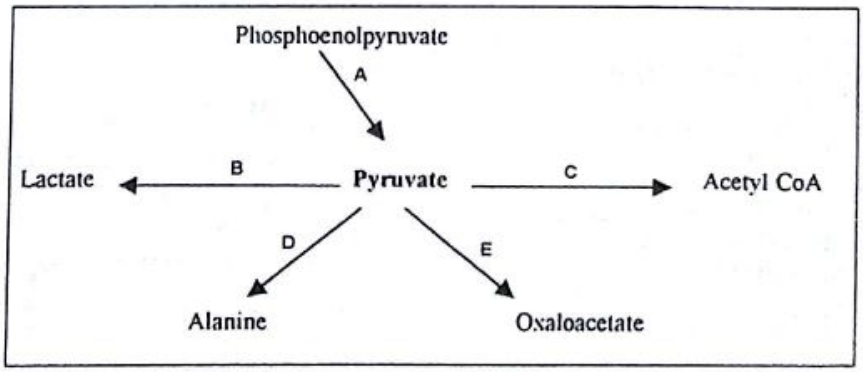
Match the numbered items and the lettered reactions as they best fit together:

298. *Requires TPP*
299. *Requires biotin*
300. *Requires pyridoxal phosphate*
301. *Regenerates oxidized NAD$^+$ in absence of O_2*
302. *Requires pyruvate kinase enzyme*



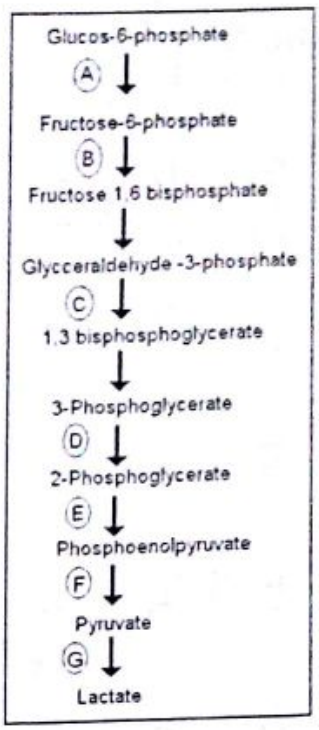
Match the numbered items and the lettered reactions as they best fit together:

303. *Requires carboxylase enzyme*
304. *Requires pyridoxal phosphate*
305. *Important for regeneration of oxidized NAD$^+$*
306. *Requires pyruvate kinase enzyme*
307. *Generates a net 3 ATP molecules*



For each reaction, select the lettered glycolytic step with which it is coupled in the diagram beside:

308. $NADH+H^+ \rightarrow NAD^+$
309. $NAD^+ \rightarrow NADH+H^+$
310. $ATP \rightarrow ADP$
311. $ADP \rightarrow ATP$
312. *Enolase*
313. *Phosphoglyceromutase*



Answer Key

MCQ:

1	2	3	4	5	6	7	8	9	10
B	D	B	D	C	B	D	D	D	A
11	12	13	14	15	16	17	18	19	20
D	D	A	D	B	E	D	C	C	E
21	22	23	24	25	26	27	28	29	30
C	C	D	A	D	C	D	C	D	D
31	32	33	34	35	36	37	38	39	40
D	A	C	B	E	C	A	C	D	D
41	42	43	44	45	46	47	48	49	50
D	C	C	E	B	A	D	E	B	B
51	52	53	54	55	56	57	58	59	60
B	B	A	A	E	B	D	C	B	C
61	62	63	64	65	66	67	68	69	70
C	B	D	B	B	A	C	D	B	D
71	72	73	74	75	76	77	78	79	80
C	C	D	C	D	B	D	D	D	B
81	82	83	84	85	86	87	88		
A	B	A	B	E	A	B	C		

True and false:

89	90	91	92	93	94	95	96	97	98
F	F	T	T	T	F	T	F	T	T
99	100	101	102	103	104	105	106	107	108
F	T	F	T	T	F	T	T	F	F
109	110	111	112	113	114	115	116	117	118
F	T	T	F	T	T	T	F	F	T
119	120	121	122	123	124	125	126	127	128
F	F	F	T	T	T	F	T	T	T
129	130	131	132	133	134	135	136	137	138
F	T	F	T	F	F	F	F	F	T
139	140	141	142	143	144	145	146	147	148
T	F	T	F	T	T	F	F	T	F
149	150	151	152	153	154	155	156	157	158
T	F	T	F	T	F	T	T	T	T
159	160	161	162	163	164	165	166	167	168
T	F	F	T	T	T	T	F	T	T
169	170	171	172	173	174	175	176	177	178
T	T	F	T	F	T	T	T	F	T
179	180	181	182	183	184	185	186	187	188
F	T	T	T	F	T	T	F	T	F
189	190	191	192	193	194	195	196	197	198
T	F	F	T	T	T	F	F	T	F
199	200	201	202	203	204	205	206	207	208
T	F	F	T	T	F	T	F	F	T
209	210	211	212	213	214	215	216	217	218
F	T	F	T	T	F	F	T	T	T
219	220	221							
T	F	F							

Matching:

222	223	224	225	226	227	228	229	230	231
B	A	D	C	D	A	C	B	C	D
232	233	234	235	236	237	238	239	240	241
B	A	B	D	A	C	B	A	D	C
242	243	244	245	246	247	248	249	250	251
C	B	D	A	D	C	A	B	B	C
252	253	254	255	256	257	258	259	260	261
A	D	E	B	E	D	A	C	D	C
262	263	264	265	266	267	268	269	270	271
A	E	B	C	A	D	B	G	F	A
272	273	274	275	276	277	278	279	280	281
B	D	B	D	A	C	D	A	C	B
282	283	284	285	286	287	288	289	290	291
E	C	D	E	A	B	E	A	B	D
292	293	294	295	296	297	298	299	300	301
C	E	C	B	A	D	C	E	D	B
302	303	304	305	306	307	308	309	310	311
A	E	D	B	A	C	G	C	B	F
312	313								
E	D								

Chapter 15 *Lipids Metabolism*

1. *Complete digestion of triacylglycerols in gastrointestinal tract needs what enzymes?*

   A. Pancreatic lipase, colipase, isomerase and bile salts.

2. *What is the function of pancreatic lipase?*

   A. Partial hydrolysis of triacylglycerols. The products are 2-monoacylglycerol and two fatty acid molecules.

3. *What is the function of isomerase?*

   A. Isomerase shifts the ester bond from position 2 to 1, this is then hydrolysed by the lipase to form free glycerol and fatty acid.

4. *Is there a complete breakdown of triacylglycerols into glycerol and fatty acids in gastrointestinal tract?*

   A. No, only partial digestion is possible.

5. *What are the final end products of digestion of triacylglycerols?*

   A. 2-monoacylglycerol (72%), 1-monoacylglycerol (6%), glycerol and fatty acids (22%).

6. *What is the function of cholesterol esterase?*

   A. It digests cholesterol esters into cholesterol and fatty acids. It also digests tibutyrin present in milk.

7. *What are types and functions of phospholipases?*

   A. They are phospholipases A_1, A_2, B and C. They digest phospholipids.

8. *How are small chain fatty acids absorbed?*

   A. Small chain fatty acids (chain length less than 12 carbons) are directly absorbed from the intestinal lumen into the portal vein and taken to the liver.

9. *How are long chain fatty acids absorbed?*

   A. Long chain fatty acids (chain length more than 12 carbons) are absorbed by forming micelles with the help of bile salts.

10. *How do bile salts help in the absorption of dietary lipids?*

A. By emulsifying the lipids and producing micelles of lipids.

11. *What is the chemical name of bile salts?*

A. Sodium glycocholate and sodium taurocholate.

12. *What are micelles?*

A. Micelles are spherical particles with a hydrophilic exterior and hydrophobic interior core. Monoacylglycerols, long chain fatty acids, cholesterol, phospholipids and lysophospholipids are incorporated into molecular aggregates to form mixed micelles.

13. *What happens to the micelles?*

A. Once inside the intestinal mucosal cell, the long chain fatty acids are re-esterified to form triacylglycerols.

14. *What is the further fate of these micelles?*

A. The triacylglycerols, together with phospholipids, and apoproteins B_{48}, are incorporated into chylomicrons. The chyle (milky fluid) from the intestinal mucosal cells loaded with chylomicrons are transported through the lacteals into the thoracic duct and then emptied into systemic circulation.

15. *What is the difference for absorption of short chain fatty acid?*

A. Short chain fatty acids are relatively soluble in water and do not need re-esterification. They directly enter into blood vessels (portal circulation).

16. *What are diet rich in short chain fatty acids?*

A. They are seen in butter, coconut oil and mother's milk.

17. *What is steatorrhea?*

A. It is abnormal increase of fat content of the stool. Normally it is less than 5 grams per day.

18. *What is steatorrhea due to?*

A. It is due to defective digestion or absorption of fat. It may also be due to diseased cells of intestinal mucosa.

19. *What are the causes of defective digestion?*

A. It is due to deficiency of pancreatic lipase, as in pancreatic duct obstruction, pancreatitis or Zollinger Ellison disease (where a high acid content of stomach enters the duodenum and inactivates lipase enzyme).

20. *What are the effects of defective digestion?*

A. If the digestion alone is defective, most of the fat excreted in feces is undigested fat. There is no loss of fat-soluble vitamins because vitamins need no digestion.

21. *What are the causes of defective absorption?*

A. Deficiency of bile salts, due to liver disease (as in cirrhosis) or bile duct obstruction (by stone, tumor of head of pancreas or enlarged lymph glands, etc.).

22. *What are the effects of defective absorption?*

A. If the absorption alone is defective, most of the fat excreted in feces is digested fat, i.e. fatty acids and monoacylglycerols. There is also loss of fat-soluble vitamins because vitamins need bile salts for absorption.

23. *What are the causes of defective cells of intestinal mucosa?*

A. It may be due to diseases in intestinal mucosa, e.g. celiac disease, sprue, Crohn's disease.

24. *What is the line of management in defective absorption?*

A. In such cases, triacylglycerols with short chain fatty acids are digested and absorbed properly, because they do not require micellerisation for absorption. Since milk fat and coconut oil are made up of short chain fatty acids, they are therapeutically useful in malabsorption syndromes.

25. *What are the functions of bile salts?*

A. Digestion of lipids (through emulsification of fat and activation of pancreatic lipase), and absorption of lipids (through combination with digested lipids to form micelles, which is water soluble and easily absorbed).

26. *What is entero hepatic circulation of bile salts:*

A. The bile salts in the intestinal lumen combine with digested lipids to form micelles, which enter the mucosa. In the mucosa, splitting of bile salts occurs. The bile salts are then reabsorbed to the liver again by blood stream. In the liver, bile salts are excreted by bile juice to reach the intestine again.

27. *White adipose tissue is concerned with what?*

A. Energy storage.

28. *Brown adipose tissue is involved in what process?*

A. Thermogenesis.

29. *What are the functions of adipose tissue (depot fat)?*

A. Energy production, fixation of some organs as kidney, Heat insulator around the body, production of vitamin D_3.

30. *How can depot fat provide energy?*

A. During fasting, the triacylglycerols stored in depot fat provide the body with free fatty acids → Acetyl CoA → Tricarboxylic acid

cycle → give energy.

31. *How can depot fat provide D_3?*

A. Exposure of skin to ultraviolet rays of sun → 7-dehydrocholesterol (present in depot fat) → vitamin D_3.

32. *What are sources of glycerol phosphate:*

A. Glycolysis (dihydroxyacetone phosphate) and glycerol (by glycerokinase → Glycerol phosphate).

33. *What are glycerol phosphate used for (fate)?*

A. Synthesis of glucose and triacylglycerols and oxidation in glycolysis.

34. *In adipose tissue, what is the source of glycerol phosphate for triacylglycerol formation?*

A. From dihydroxyacetone phosphate, derived from glucose.

35. *Triacylglycerol synthesis is enhanced by which hormone?*

A. Insulin.

36. *What enzymes hydrolyze triacylglycerols present in adipose tissue?*

A. Hormone sensitive triacylglycerols lipase, diacylglycerol lipase and monoacylglycerol lipase.

37. *What key enzyme is involved in lipolysis?*

A. Hormone sensitive triacylglycerols lipase.

38. *Which hormones are the lipolytic?*

A. Glucagon, adrenalin, growth hormone, corticosteroids and ACTH.

39. *What is the mechanism of activation of hormone sensitive lipase?*

A. Through activation of adenylate cyclase, cyclic AMP and protein kinase.

40. *Which hormones inhibit hormone sensitive lipase?*

A. Insulin.

41. *What is the mechanism of inhibition of hormone sensitive lipase?*

A. (1) Through activation of phosphodiesterase enzyme → ↓ cAMP → inhibition of lipolysis. (2) Insulin also stimulates phosphatase enzyme which dephosphorylates hormone sensitive lipase → inhibition of lipolysis

42. *Does caffeine favor obesity or loss of weight?*

A. Caffeine is a substance present in coffee and tea. It inhibits phosphodiesterase enzyme → Stimulation of lipolysis. Thus, it favors loss of weight.

43. *What are types of fatty acids oxidation:*

A. α, β and ω oxidation.

44. *How fatty acids are activated in preparation of oxidation?*

A. Fatty acids are activated to their co-enzyme A (CoA) derivative i.e. acyl CoA.

45. *What is the enzyme for this activation?*

A. Fatty acyl CoA synthetase.

46. *How much ATP is required for this reaction?*

A. One molecule of ATP is hydrolysed to AMP and PPI. Thus, two high energy bonds are utilized in this reaction.

47. *What are the coenzymes needed for fatty acid oxidation?*

A. FAD and NAD.

48. *What is carnitine?*

A. Carnitine is β-hydroxy-δ-trimethyl ammonium butyrate. It is synthesized from lysine and methionine in liver and kidney.

49. *What is the function of carnitine?*

A. Carnitine is involved in transfer of long chain fatty acids to inside the mitochondria.

50. *What about short chain fatty acids?*

A. Short chain fatty acids do not require carnitine for transport, so they are easily oxidized.

51. *What is the net generation of ATP, when one molecule of palmitic acid (16 carbon) is oxidized completely?*

A. 129.

52. *How?*

A. β-Oxidation of palmitic acid will be repeated 7 times (turns) to produce 8 acetyl CoA. In each turn, one molecule of reduced $FADH_2$ and one molecule of reduced $NADH+H^+$ are produced. They are oxidized in respiratory chain to give 5 ATP. (FADH2 → 2 ATP) and ($NADH+H^+$ → 3 ATP) ∴ 7 turns × 5 ATP → 35 ATP. Oxidation of one molecule of acetyl CoA in citric acid cycle gives 12 ATP. ∴8 Acetyl CoA × 12 ATP = 96 ATP. Two high energy phosphate bonds are utilized in the first reaction (catalyzed by acyl CoA synthetase) which occurs for one time only. ∴ Net energy gain = Energy produced – Energy utilized =(35 ATP + 96 ATP) – 2 ATP = 131 ATP – 2 ATP = 129 ATP

53. *What are the products during each cycle of β-oxidation of fatty acid?*

A. Acetyl CoA, FADH2, and $NADH+H^+$.

54. *What are the energy producing steps in β-oxidation pathway?*

A. Fatty acyl CoA dehydrogenase (FAD) and β-hydroxy fatty acyl CoA dehydrogenase (NAD) steps.

55. *What is the product of β-oxidation of odd chain fatty acids?*

A. Propionyl CoA.

56. *What is the further metabolism of propionyl CoA?*

A. Propionyl CoA is first carboxylated to methyl malonyl CoA and then to succinyl CoA.

57. *What are the co-enzymes required for the conversion of propionyl CoA to succinyl CoA?*

A. Biotin, ATP, Vitamin B12.

58. *Succinyl CoA is generated from which substances?*

A. Odd chain fatty acids, propionic acid, citric acid cycle and some amino acids as valine, isoleucine.

59. *Succinyl CoA is utilised for what purposes?*

A. Heme synthesis, activation of acetoacetate, oxidation in TCA cycle, glucose synthesis and detoxication.

60. *What is alpha oxidation of fatty acid?*

A. It is a process by which fatty acids are oxidized by removing alpha carbon atoms, one at a time. It is used for fatty acids that have a methyl group at the beta-carbon, which blocks beta-oxidation. Alpha oxidation does not generate energy.

61. *Where is alpha oxidation taking place?*

A. In endoplasmic reticulum (microsomes).

62. *Refsum's disease is due to what?*

A. Accumulation of phytanic acid, due to defective alpha oxidation.

63. *When unsaturated fatty acids are oxidised, how many ATP is formed?*

A. The energy yield is less by 2 ATP molecules per double bond, when compared to the corresponding chain length saturated fatty acid.

64. *Why 2 ATP molecules are reduced?*

A. Because, the FAD dependent dehydrogenation (step 1 of beta oxidation) does not occur at the double bond.

65. *When palmitoleic acid (16 C, 1 double bond) is completely oxidised, what is the net generation of ATP molecules?*

A. 127.

66. *What are the sources of acetyl CoA?*

A. Carbohydrates (Pyruvate), lipids (fatty acids & acetoacetyl CoA), and proteins (as leucine).

67. *What are the fate of acetyl-CoA?*

A. Fatty acid synthesis, oxidation in citric acid cycle, cholesterol synthesis, ketone body synthesis and acetyl choline synthesis.

68. *What are the types of fatty acids synthesis.*

A. Three mechanisms, cytoplasmic, mitochondrial and microsomal.

69. *What are the sources of NADPH+H$^+$?*

A. Pentose phosphate pathway, action of malic enzyme on malate and action of cytosolic isocitrate dehydrogenase on isocitrate.

70. *What are the enzymes used for synthesis of NADPH for fatty acid synthesis?*

A. Glucose-6-phosphate dehydrogenase, malic enzyme and cytosolic isocitrate dehydrogenase.

71. *Which step of pentose phosphate pathway produces NADPH+H$^+$?*

A. The first reaction catalyzed by glucose-6-phosphate dehydrogenase.

72. *What is the rate limiting enzyme of de novo synthesis of fatty acid?*

A. Acetyl CoA carboxylase.

73. *What is the reaction catalyzed by acetyl CoA carboxylase?*

A. Acetyl CoA + CO2 → Malonyl CoA.

74. *What are the co-enzymes required for the reaction?*

A. Biotin and ATP.

75. *What is the fatty acid synthase complex:*

A. This enzyme is a dimer i.e. formed of 2 subunits. Each unit, (monomer), contains 7 enzymes and a terminal protein called acyl carrier protein (ACP).

76. *Acetyl CoA from mitochondria is transferred to cytosol for the de novo synthesis of fatty acid, by which enzyme?*

A. ATP citrate lyase.

77. *How can acetyl CoA get its way to cytoplasm for palmitic acid synthesis?*

A. Acetyl CoA condenses with oxaloacetate – in the presence of citrate synthase- to form citrate. Then citrate diffuses out of mitochondria to cytosol where it is spit again – by citrate lyase – into acetyl CoA and oxaloacetate.

78. *What are the steps in which NADPH is used in fatty acid synthesis?*

A. Step 4 (Ketoacyl reductase) and step 6 (Enoyl reductase).

79. *Explain why the acetyl CoA derived from oxidation of glucose and not from fatty acid is the only source for fatty acid synthesis?*

A. This because insulin hormone secreted after meal stimulates both

glucose oxidation (→ acetyl CoA) and lipogenesis (=Fatty acid synthesis) and inhibits lipolysis.

80. *How is fatty acid synthesis regulated?*

A. Key enzyme, acetyl CoA carboxylase, is stimulated by insulin and citrate and inhibited by long chain acyl CoA (palmitoyl CoA).

81. *What is the action of insulin on fatty acid synthesis?*

A. Insulin favors lipogenesis.

82. *How?*

A. Insulin enhances the uptake of glucose by adipocytes and increases the activity of pyruvate dehydrogenase. So, availability of acetyl CoA is increased. Insulin also activates glucose-6-phosphate dehydrogenase, so that enough NADPH is available. Moreover, insulin stimulates acetyl CoA carboxylase, the key enzyme of fatty acid synthesis pathway. Insulin also depresses the hormone sensitive lipase (inhibits lipolysis).

83. *Chain elongation of fatty acid is taking place in which site?*

A. Microsomal elongation system is more active.

84. *Where does desaturation of fatty acid takes place?*

A. In the endoplasmic reticulum.

85. *What is the enzyme called?*

A. Microsomal desaturase system.

86. *What are essential fatty acids?*

A. Those cannot be synthesized by the body. So they are to be provided in the diet.

87. *Name the essential fatty acids.*

A. Linoleic and linolenic acids are the only fatty acids, which cannot be synthesized in the body.

88. *What are the important substances derived from PUFA?*

A. Prostaglandins, prostacycline, thromboxanes, leukotrienes, and HPETE.

89. *What are the functions of PUFA?*

A. Synthesis of prostaglandins, synthesis of phospholipids and esterification of cholesterol.

90. *Enumerate eicosanoids* :

A. They include prostanoids (prostaglandins [PGE_2 and $PGF_2\alpha$], Prostacyclin [PGI_2], and thromboxane A_2 [TXA_2]) and leukotriene A_4 [LTA], LTC_4, LTD_4 and LTD_4, LTB_4.

91.How prostaglandins are classified?

A. According to the attachment of different substitute groups to the ring, PGs are named with capital letters such as A,B,E and F. In the same series, depending on number of double bonds on the side chains they are denoted by a subscript after the capital letter, e.g.PGE1, PGE2, PGE3,etc. Series 2 have two double bonds at 13-14 (trans) and 5-6 (cis). This is the most common variety.

92.Prostaglandins are derived from what substance?

A. Prostaglandins are derived from the PUFA. The series 2 (with two double bonds) are derived from arachidonic acid. All naturally occurring PGs belong to the 2-series.

93.Prostaglandins are stored in what form?

A. As precursors, as membrane phospholipids.

94.How prostaglandins are synthesized?

A. The arachidonic acid is released by the action of phospholipase A_2 on phospholipids. Prostaglandins synthesis is catalyzed by prostaglandin H synthase (PGHS) that contains two separate enzyme activities, cyclo-oxygenase and peroxidase.

95.How prostaglandins synthesis is regulated?

A. The phospholipase is activated by epinephrine. Steroids inhibit PL and prevent release of arachidonic acid from membranes.

96.What is the importance of cyclo-oxygenase?

A. Cyclo-oxygenase is activated by catecholamines and inhibited by non-steroid anti-inflammatory drugs (NSAIDS). Cyclo-oxygenase is a "suicide" enzyme.

97.What is the mechanism of action of aspirin?

A. Aspirin acetylates serine in the active site and irreversibly inhibits the cyclo-oxygenase.

98.How is prostaglandins inactivated?

A. Prostaglandins have only very short half life, of about 30 seconds. They are inactivated by the 15-hydroxy-prostaglandin-dehydrogenase, which converts 15-OH group to keto group.

99.What is the mechanism of action of prostaglandins?

A. Prostaglandins are local hormones, and function through G-protein coupled receptors in most tissues, PGE increases cAMP level. But in adipose tissue and in renal tubular cells, PGE lowers cAMP level.

100. *What is the action of prostacyclin on vascular endothelium?*

A. Prostacyclin causes vasodilataion. It also inhibits platelet aggregation and has protective effect on vessel wall against deposition of platelets.

101. *What is the effect of thromboxane?*

A. Thromboxane (TXA2) is the main PG produced by platelets. The major effects are vasoconstriction and platelet aggregation. Prostacyclin and thromboxane are opposing in activity.

102. *what is the precursor of leukotrienes?*

A. They are produced from arachidonic acid.

103. *What is its biological importance?*

A. LT B4 is produced in neutrophils, it is the most potent chemotactic agent. The slow reacting substance of anaphylaxis (SRS-A) contains LTC4, LTD4, LTE4. They cause smooth muscle contraction, constrict the bronchioles, increase capillary permeability, and produce vasoconstriction.

104. *How phospholipids are classified?*

A. Glycerophospholipids (containing glycerol) and sphingophospholipids (containing sphingosine).

105. *Give example for Glycerophospholipids*

A. Lecithin, cephalin, cardiolipin and plasmalogens.

106. *Give example for sphingophospholipids*

A. Sphingomyelin.

107. *Name some lipid storage diseases.*

A. Tay Sach's disease, Niemann Pick's disease, Gaucher's disease.

108. *Gaucher's disease is due to the deficiency of what?*

A. Beta glucosidase *(glucocerebrosidase)*.

109. *What is accumulated in Gaucher's disease?*

A. Glucocerebroside.

110. *What is the manifestations of Gaucher's disease?*

A. Enlarged liver and spleen and mental retardation.

111. *Niemann-Pick disease is due to the deficiency of what?*

A. Sphingomyelinase.

112. *What is accumulated in Niemann-Pick disease?*

A. Sphingomyelin.

113. *What is the manifestations of Niemann-Pick disease?*

A. Enlarged liver and spleen due to accumulation of sphingomyelin; fatal in early life.

114. *Tay Sach's disease is due to the deficiency of what?*

A. Hexosaminidase.

115. *What is accumulated in Tay Sach's disease?*

A. Ganglioside.

116. *What are ketone bodies?*

A. Aceto acetate, β-hydroxy butyric acid, and acetone.

117. *What are the Functions of ketone bodies?*

A. Energy production for most tissues. acetoacetyl CoA is split into 2 acetyl CoA, each gives 12 ATP in TCA.

118. *Can liver utilize ketone bodies?*

A. NO because liver does not contain enzymes for ketone bodies oxidation (ketolysis). Thus, liver cannot oxidize them.

119. *Can brain cells oxidize ketone bodies?*

A. Yes, after 5-6 days of fasting

120. *Ketone bodies are formed in which tissue?*

A. Liver mitochondria.

121. *What is the rate-limiting-step in ketone body formation?*

A. HMGCoA synthase.

122. *HMG CoA is directly converted into what substances?*

A. Acetacetate, acetyl CoA and mevalonate.

123. *Ketone body utilisation is taking place in which organs?*

A. Ketolysis is taking place in extra hepatic tissues (All other tissues, except liver).

124. *Utilisation of ketone bodies by peripheral tissues needs which enzyme?*

A. Succinyl CoA thiophorase.

125. *What is the concentration of ketone bodies?*

A. Blood ketone bodies concentration is less than 3 mg/dl.

126. *What is ketonemia?*

A. It is the increase of blood ketone bodies above normal concentration.

127. *Ketonemia is due to what processes?*

A. Ketonemia occurs when the rate of formation of ketone bodies ketogenesis) is grater than the rate of their oxidation (ketolysis). Step 1: Absence of insulin leads to excessive lipolysis. Step 2: So more fatty acid is available, and more acetyl CoA is produced. Step 3: But oxidation of acetyl CoA in citric acid cycle is sluggish. The excess acetyl CoA is derived into ketone body formation.

128. *What is ketonuria?*

A. It is the increase of urine ketone bodies concentration above normal concentration. It usually occurs with ketonemia.

129. *What test is used to identify ketone bodies in urine?*

A. Rothera's test.

130. *What is ketosis:*

A. It is a condition of metabolic acidosis results from ketonemia.

131. *What is the mechanism of ketosis?*

A. Increase ketone bodies in blood is neutralized by blood buffers mainly bicarbonate (HCO_3^-). Bicarbonate will be depleted and this leads to decreased blood pH (acidosis).

132. *What are the effects of ketosis?*

A. Dizziness, loss of concentration, hyperkalemia. In severe cases coma may develop.

133. *What are conditions in which ketonemia developed?*

A. Starvation, severe diabetes mellitus, hyper-catabolic states e.g. diarrhea and fever and tuberculosis.

134. *What is the ring structure present in cholesterol?*

A. Cyclopentano perhydro phenanthrene ring (steroid ring).

135. *Cholesterol has how many carbon atoms?*

A. 27.

136. *What are the substances derived from cholesterol?*

A. Vitamin D3, bile salts, and steroid hormones (Glucocorticoids, mineralocorticoids, testosterone, estrogens and progesterone).

137. *What are the functions of cholesterol?*

A. Cholesterol is a constituent of cell membrane. Cholesterol is also the precursor of vitamin D_3, bile salts, steroid hormones.

138. *Which food stuffs contain cholesterol?*

A. Non-vegetarian food as meat and liver.

139. *is cholesterol present in vegetable oils?*

A. No.

140. *What is the rate-limiting-step (key enzyme) in the cholesterol biosynthesis?*

A. HMG CoA reductase.

141. *How can cholesterol synthesis be regulated?*

A. HMG CoA reductase is regulated through feed back inhibition, feed back regulation, hormonal regulation and inhibition by drugs.

142. *How can cholesterol synthesis be regulated by feed back inhibition?*

A. Cholesterol (the end product of the pathway) acts as feed back inhibitor of HMG CoA reductase enzyme. Thus, it decreases more cholesterol synthesis.

143. *How can cholesterol synthesis be regulated by feed back regulation?*

A. Cholesterol (either synthesized by the cell or reaching it from diet) inhibits HMG CoA reductase gene. This decreases transcription and synthesis of HMG CoA reductase.

144. *What are hormones regulating cholesterol synthesis?*

A. Glucagon Inhibits HMG CoA reductase and Insulin stimulates HMG CoA reductase.

145. *What are drugs regulating cholesterol synthesis?*

A. Lovastatin and mevastatin are drugs, which inhibit HMG CoA reductase by reversible competitive inhibition.

146. *What are sources of HMG CoA:*

A. Acetyl CoA. It is condensed with acetoacetyl CoA (in the presence of HMG CoA synthase) to form HMG CoA.

147. *What are the fate of HMG CoA:*

A. Ketone bodies formation (in the mitochondria) and mevalonate and cholesterol formation (in cytosol).

148. *What is the first sterol ring formed during cholesterol biosynthesis?*

A. Lanosterol.

149. *How cholesterol is excreted?*

A. Through bile. About one gram of cholesterol is excreted daily partly as cholesterol itself, and partly as bile salts.

150. *What is the normal level of total plasma choesterol?*

A. 140-220 mg/dl.

151. *What is hyper-cholesterolemia?*

A. It is increased plasma cholesterol concentration above 220 mg/dl.

152. *What are causes of hypercholesterolemia?*

A. Diabetes mellitus, nephrotic syndrome, obstructive jaundice, hypothyroidism and familial hypercholesterolemia.

153. *How does diabetes mellitus cause hypercholesterolemia?*

A. Insulin stimulates lipoprotein lipase that clear plasma lipoproteins and cholesterol. Insulin is deficient in diabetes mellitus.

154. *How does hyporthyroidism cause hypercholesterolemia?*

A. Thyroid hormones stimulate conversion of cholesterol to bile acids.

155. *What is the hypo-cholesterolemia?*

A. It is decreased plasma cholesterol concentration below 140 mg/dl.

156. *What are the causes of hypo-cholesterolemia?*

A. Prolonged fasting due to decreased activation of HMG-CoA reductase, liver diseases, where most plasma cholesterol is synthesized, hyperthyroidism and chronic infection as tuberculosis. Diet rich in unsaturated fatty acids and poor in saturated fatty acids, carbohydrate and cholesterol causes of hypo-cholesterolemia.

157. *What advise will you give to a person with increased cholesterol level?*

A. Reduce food with higher cholesterol content, include PUFA and omega-3 fatty acids in diet, reduction of total fat intake of green leafy vegetables, exercise, and avoid cigarette smoking.

158. *When will you start drugs for a person with increased cholesterol level?*

A. When patient's condition is not responding to the dietary restriction.

159. *What are the drugs available to treat hypercholesterolemia?*

A. HMG CoA reductase inhibitors (lovastatin and mevastatin), nictotinic acid and bile acid binding resins,.

160. *What is the function of bile acids?*

A. They are the major route of excretion of cholesterol and they are required for absorption of triacylglycerols.

161. *Bile acids are derived from what substance?*

A. Cholesterol.

162. *How are bile salts formed?*

A. Bile acids are conjugated with taurine or glycine.

163. *What is cholesterol precursor for?*

A. Bile acids, vitamin D_3, Steroid hormones.

164. *What are steroid hormones?*

A. Glucocorticoids, mineralocorticoids, male sex hormones (testosterone) and female sex hormones (estrogens and progesterone). Also active vitamin D_3 (calcitriol) is a steroid hormone.

165. *What is the normal level of total plasma lipids?*

A. 360-820 mg/dl.

166. *What is the normal level of triacylglycerols?*

A. 40-160 mg/dl.

167. *What are types of apolipoproteins?*

A. Apo. A (I, II and IV), apo. B (48 and 100), apo. C (I,II and III) , apo. D and apo. E.

168. *What are the functions of apolipoproteins?*

A. (1) Apolipoproteins combine with water insoluble lipids to form water soluble lipoproteins. This helps transport of lipids between tissues in aqueous plasma. (2) Apolipoproteins activate some enzymes e.g. apo C-II activates lipoprotein lipase enzyme (3) Apolipoproteins act as ligands that bind with lipoprotein receptors in tissues e.g. apo B_{100} and apo E for LDLreceptors.

169. *How lipoproteins are estimated?*

A. Either by electrophoresis or by ultracentrifugation.

170. *During electrophoresis, what is the fastest moving lipoprotein?*

A. HDL (α-lipoprotein).

171. *During electrophoresis, what is the least moving lipoprotein?*

A. Chylomicron (δ-position).

172. *Maximum triacylglycerols content is in which lipoprotein?*

A. Chylomicrons.

173. *Maximum cholesterol content is in which lipoprotein?*

A. LDL (β position).

174. *Triacylglycerols present in chylomicrons are hydrolysed by what?*

A. Lipoprotein lipase.

175. *Where is the enzyme present?*

A. It is located at the endothelial layer of capillaries of adipose tissue, muscles, and heart, but not in liver.

176. *What is the main apoprotein present in chylomicrons?*

A. B48.

177. *Where is apo B_{48} synthesized?*

A. Intestinal mucosal cells.

178. Why it is named apo B48?

A. Because its molecular weight is 48% of the molecular weight of B100.

179. *What is the function of chylomicrons?*

A. Transport of dietary lipids (mainly triacylglycerols) from intestine to adipose tissue.

180. *Where are chylomicrons synthesized?*

A. Intestinal mucosal cells.

181. *What is the structure of chylomicrons?*

A. Main lipids are triacylglycerols. They contain also cholesterol, phospholipids and fat-soluble vitamins. Proteins are (2%), apo B_{48} and receives apo CII and apo E from HDL.

182. *What is the fate (catabolism) of Chylomicrons?*

A. TG are hydrolyzed by lipoprotein lipase (which is activated by apo CII). The remaining parts are chylomicron remnants, which are then taken up by the liver. Hepatocyte receptors can recognize apoB_{48} and apo E.

183. *Endogenous triacylglycerols in plasma are carried by what?*

A. VLDL.

184. *What is the main apoprotein present in VLDL?*

A. B100.

185. *Where is apo B100 synthesized?*

A. Liver.

186. *What is the function of VLDL?*

A. Transport lipids mainly triacylglycerols from liver to peripheral tissues.

187. *Where are VLDL synthesized?*

A. Liver.

188. *What is the structure of VLDL?*

A. Main lipids: triacylglycerols. contains also cholesterol, phospholipids. Proteins contents (10%) include apo B_{100} and receive apo CII and apo E from HDL.

189. *What is the fate (catabolism) of VLDL?*

A. Triacyglycerols are hydrolyzed by lipoprotein lipase. The remaining parts are IDL, which are then converted into LDL by transferring phospholipids, apo C and apo E to HDL.

190. *What is the main apoprotein present in LDL?*

A. B100, it is the ligand for LDL receptor.

191. *What is the function of LDL?*

A. Transport of cholesterol from liver to peripheral tissues.

192. *Where are LDL synthesized?*

A. Circulation from VLDL and IDL.

193. *What is the structure of LDL?*

A. Lipid contents: cholesterol, cholesterol esters and phospholipids. Protein contents: (22%), apo B_{100}.

194. *What is the fate (catabolism) of LDL?*

A. LDL apo B_{100} are recognized by tissue receptors. After binding with receptors, the LDL are internalized by endocytosis. Inside cells LDL are separated from receptors and hydrolyzed by lysosomal enzymes releasing cholesterol, amino acids, fatty acids and phospholipids.

195. *What is the function of LDL receptors?*

A. LDL receptors are negatively charged glycoproteins present on all cells but most abundant in hepatic cells and adrenal cortex. LDL receptors, located in specialized regions called clathrin-coated pits. When the apo B-100 binds to the receptor, the receptor-LDL complex is internalized by endocytosis.

196. *What is the fate of cholesterol inside liver cells?*

A. Step-1: Forms cholesterol ester by ACAT enzyme. Step-2: Inhibits HMG CoA reductase. This inhibits cholesterol synthesis by the cell. Step-3: inhibits DNA synthesis of LDL receptors. This leads to decrease LDL receptors (down regulation) and inhibition of LDL uptake by cells.

197. *What is the function of HDL?*

A. Transport of cholesterol from peripheral tissues to liver.

198. *How is this function achieved?*

A. HDL contains Apo A-1, which activates LCAT enzyme, which forms cholesterol ester. This helps to remove cholesterol from peripheral tissues to the liver. It contains also Apo C-II, which activates lipoprotein lipase for hydrolysis of triacylglycerols.

199. *What is the main apoprotein present in HDL?*

A. Apo A-1, it is the ligand for HDL receptor.

200. *What is LCAT?*

A. Lecithin cholesterol acyl transferase enzyme.

201. *Where is it present?*

A. LCAT is present in plasma and activated by apo-A1, when LACT binds to HDL disc.

202. What is the importance of LCAT?

A. The free cholesterol is esterified by LCAT. The esterified cholesterol is then incorporated into HDL disc, to form mature HDL. So for excretion of cholesterol, LCAT is necessary.

203. What is the structure of HDL?

A. Main lipids cholesterol (free and esterified) and phospholipids, mainly lecithin. Proteins contents (50%) include apo A-1 (which activates LCAT), apo C (which activates lipoprotein lipase and E (which is recognized by hepatic receptors)

204. What is the fate (catabolism) of HDL?

A. HDL accepts unesterified cholesterol from peripheral tissues. Then HDL (by LCAT enzyme) esterifies cholesterol into cholesterol esters. Then HDL is taken up by the liver cells → where cholesterol esters are released inside these cells → to be utilized in the formation of lipoproteins or excreted in bile.

205. What is "bad cholesterol"?

A. LDL cholesterol.

206. Why is it called so?

A. LDL transportes cholesterol from liver to peripheral tissues, where it is deposited, and causes atherosclerosis.

207. What is lipoprotein (a)?

A. It is attached to apo B-100 by a disulfide bond. It has significant homology with plasminogen. So it interferes with plasminogen activation and impairs fibrinolysis. This leads to unopposed intravascular thrombosis and possible myocardial infraction.

208. What is the significance of lipoprotein (a)?

A. Lp(a) is associated with heart attacks at the age of 30 or 40 years. Indians have a higher level of Lp(a) than Europeans.

209. What is the normal level of lipoprotein (a)?

A. In 40% population, there is no detectable level of Lp(a) in serum. In 20% of population, the Lp(a) concentration in blood is more than 30 mg/dl, and these persons are susceptible to heart attack at a younger age.

210. What is "good cholesterol"?

A. HDL cholesterol.

211. Why is it called so?

A. HDL transportes cholesterol from peripheral tissues to liver, and so helps in excretion of cholesterol from the body. So HDL is antiatherogenic.

212. *Free fatty acids of plasma are bound to what?*

A. Bound to serum albumin.

213. *What is the function of albumin?*

A. Transport of free fatty acid from adipose tissue to peripheral tissues.

214. *What are the features of hyperlipoproteinemia Type II-A?*

A. Atherosclerosis and elevated plasma LDL cholesterol

215. *What is its cause?*

A. Defect in LDL receptor.

216. *How atherosclerosis started?*

A. The effect is directly proportional to the LDL level. Free radical induced oxidative damage of LDL will accelerate this process. Oxidised LDL cholesterol is deposited in the subintimal regions of arteries.

217. *What vessels are affected mostly by atherosclerosis?*

A. Aorta, coronary arteries and cerebral vessels are predominantly affected.

218. *How LDL deposit leads to atherosclerosis?*

A. Oxidised LDL is taken up by macrophages, the macrophages become over-loaded with cholesterol esters, and these are then called "foam cells" which form the hallmark of atherosclerotic plaques.

219. *What happens to atherosclerotic plaque?*

A. This leads to narrowing of vessel wall. Then fibrous proliferation takes place, this is due to liberation of various growth factors. Again a clot is formed which occludes one of the major vessels. Then there is ischemia of the tissue supplied. Finally infarction or ischemic death of the tissue occurs.

220. *What are the important risk factors of coronary artery diseases?*

A. Serum cholesterol level above 220 mg/dl, LDL-cholesterol level above 160 mg/dl, HDL-cholesterol level below 35 mg/dl, and Apo(a) above 30 mg/dl.

221. *What are other risk factors associated with coronary artery diseases?*

A. Cigarette smoking, hypertension, diabetes mellitus, serum triglyceride level above 200 mg/dl, homocysteine level, sedentary life style, obesity.

222. *Primary hyperlipoproteinemia (Type I).is due to the deficiency of what?*

A. Lipoprotein lipase.

223. *What is accumulated in primary hyperlipoproteinemia?*

A. Chylomicrons and VLDL.

224. *In the blood, fatty acids are transported as what form?*

A. Albumin is the carrier of free fatty acid.

225. *What are polyunsaturated fatty acids (PUFA)?*

A. These are fatty acids that contain more than one double bond e.g. lenolenic acid and arachidonic acid.

226. *What is the importance of polyunsaturated fatty acids (PUFA) in cholesterol metabolism?*

A. PUFA present in lecithin is transferred to cholesterol by the enzymes LACT. The esterified cholesterol is then taken by HDL, and finally excreted through liver. So, for excretion of cholesterol, PUFA is required. Thus, PUFA will lower the blood level of cholesterol.

227. *What are lipotropic factors?*

A. Lipotropic factors are substances that help the mobilization of fat from the liver i.e. prevent fatty liver.

228. *What are types of lipotropic factors?*

A. phospholipids, amino acids (as methionine & serine), vitamins (as vitamin B_{12} & folic acid), proteins of high biological value.

229. *Give examples of substances that prevent fatty liver?*

A. Lecithin, choline and methionine.

230. *Why are phospholipids considered as lipotropic factors?*

A. Because they are relatively soluble in water.

231. *What are amino acids acting as lipotropic factors?*

A. Methionine, which is a methyl donor essential for choline formation and serine, which enters in structure of cephaline.

232. *What are vitamins acting as lipotropic factors?*

A. Vitamin B_{12} and folic acid have a role in synthesis and transfer of methyl group (CH_3).

233. *What worsens fatty liver?*

A. Alcohol, diabetes mellitus, excess caloric intake, hepatitis virus.

234. What are the types of lipase enzymes?

A. (1) Digestive lipases (gastric, pancreatic, intestinal and lingual lipases), (2) blood vessels lipase (lipoprotein lipase), (3) adipose tissue lipases (hormone sensitive triacylglycerols lipase, diacylglycerols lipase, and monoacylglycerols lipase), (4) Tissue lipase (acid lipase), and (5) liver lipase (hepatic lipase).

235. What is the role of liver in lipids metabolism:

A. Fatty acid synthesis and oxidation, synthesis and esterification of cholesterol, formation of lipoproteins, formation of phospholipids, formation of ketone bodies, formation of bile salts, storage of fat soluble vitamins, and detoxication of steroid hormones.

236. Compare between Fatty acid oxidation and fatty acid synthesis:

	Fatty acid oxidation	fatty acid synthesis
Site	Mitochondria	Cytosol
Enzymes	independent	grouped as a multi-enzyme complex
Coenzymes	FAD and NAD	NADPH
During the process	2 carbon units are removed as acetyl CoA	2 carbon units are added as 3 carbon Malonyl CoA

237. Compare between Apolipoproteins:

Apolipo-proteins	LIPOPROTEINS	COMMENT
Apo A(I, II and III)	HDL-chylomicrons	*Activator of LCAT *Ligands for HDL-receptors
Apo B_{-100}	LDL-VLDL-IDL	*Synthesized by liver *Ligand for LDL-receptors in tissues
Apo B_{-48}	Chylomicrons-chylomicron remnants	*Synthesized by intestine *Ligand for chylomicron remnant receptors in liver.
Apo C (I, II and III)	Chylomicrons-VLDL-HDL	Activator of lipoprotein lipase.
Apo D	HDL	May act as lipid transfer protein.
Apo E	Chylomicrons-Chylomicron remnants-VLDL-HDL	*Ligand for chylomicron remnant receptors in liver. *Ligand for LDL receptors in tissues.

238. Compare between hyperlipoproteinemias:

Types	↑ Plasma Lipoproteins	↑ Blood lipids	Atherosclerosis
Type I	Chylomicrons	Triacylglycerols	Not liable
Type II	LDL	Cholesterol	Very liable
Type III	VLDL & LDL	Triacylglycerols	Liable
Type IV	VLDL	Triacylglycerols	Not liable
Type V	Chylomicrons& VLDL	Triacylglycerols	Not liable

239. Compare between Fatty acid synthesis.

	Cytoplasmic	Microsomal	Mitochondrial
Site	Cytosol	Microsomes	Mitochondria
Nature	Synthesis of palmitate	Elongation of preexisting fatty acid	Reverse of β-oxidation
ACP	Needed	Not needed	Not needed
Source of 2 carbons	Malonyl CoA	Malonyl CoA	Acetyl CoA
Coenzymes	$NADPH+H^+$	$NADPH+H^+$	$NADPH+H^+$ and $NADH+H^+$

240. Compare between tissue fat and depot fat

	Tissue fat	Depot fat
Site	Cell membrane	Adipose tissue
Amount	Limited	Unlimited
Composition	Mainly phospholipids	Mainly triacylglycerols
Oxidation	Never oxidized to give energy	Used as a source of energy
Functions	Responsible for selective permeability	Source of energy – Fixation of organs – Vitamin D_3 – Heat insulator.

241. Compare between lipase enzymes:

Enzyme	Origin	Site of action	Function	Special properties
Gastric lipase	Stomach	Stomach	Degrades dietary triacylglycerols in infants	Needs acid pH
Pancreatic lipase	Pancreas	Small intestine	Degrades dietary triacylglycerols (removes fatty acid from carbon 1 and 3, leaving 2-monoglycerol)	Needs co-lipase
Lipoprotein lipase	Blood and extra-hepatic tissues	Surface endothelial cells lining the capillaries	Degradeds TG circulating in chylomicrons or VLDL, releasing FA and glycerol	Can be released into plasma by heparin; activated by apo. C-II
Hormone sensitive lipase	Adipose tissue	Adipose tissue	Degradation of stored triacylglycerols	Activated by cAMP
Acid lipase	Most tissues	Lysosomes	Removes fatty acids from lipids taken into cells during phagocytosis	Needs acid pH
Hepatic lipase	Liver	Liver	Removes fatty acids from chylomicron remnants and HDL taken by hepatic cells.	

242. Compare between cholesterol synthesis and ketone bodies synthesis:

	cholesterol synthesis	ketone bodies synthesis
Building blocks	Acetyl CoA derived from glucose oxidation	Acetyl CoA derived from fatty acids (lipolysis).
Site of synthesis	Cytosol	Mitochondria
	Liver, subcutaneous fat, adrenal cortex and gonads.	Liver only

Insulin	Stimulates synthesis	Inhibits synthesis
HMG CoA	Gives mevalonate → Cholesterol	Gives acetoacetate (ketone bodies)

243. *Compare between HDL and LDL*

	HDL	LDL
Synthesis	Liver	Circulation (from VLDL)
Functions	*Remove cholesterol to peripheral tissues. *Contain Apo CII → Activate lipoprotein lipase → Hydrolysis of TG	Provide cholesterol to peripheral tissues.
Composition	Lower concentration of cholesterol and cholesterol esters.	Higher concentration of cholesterol and cholesterol esters.
Protein content	*Apo A-I → Stimulates LCAT *Apo C-II → Stimulates lipoprotein lipase	Apo B_{48}, which is recognized by hepatic cells.
Catabolism	HDL transfers Cholesterol (after esterified) from peripheral tissues to the liver.	LDL is internalized by the cells and stored in the form of cholesterol esters.

What is the catalytic activity, intracellular location and pathway of the following enzymes?

244. *Hormone sensitive triacylglycerol lipase*
245. *HMG CoA reductase.*
246. *HMG CoA lyase.*
247. *Acetyl CoA carboxylase*
248. *ACAT*
249. *LCAT*
250. *Pancreatic lipase*
251. *Carnitine palmitoyl-transferase II*
252. *Fatty acid cyclooxygenase*

Enzyme	*Catalytic activity*	*Location*	*Pathway*
Hormone sensitive TG lipase	TG → Glycerol + FA	Cytosol	Lipolysis
HMG CoA reductase	HMG → Mevalonate	Cytosol	Cholesterol synthesis
HMG CoA lyase	HMG → Acetoacetate	Mitochondria	Ketogenesis
Acetyl CoA carboxylase	Acetyl CoA → Malonyl CoA	Cytosol	Cytoplasmic FA synthesis
ACAT	Acyl CoA + Cholesterol → Cholesterol ester + CoA	Liver	Catabolism of LDL
LCAT	Cholesterol + Lecithin → Cholesterol ester + Lysolecithin	Cytosol	Catabolism of HDL
Pancreatic lipase	TG → Monoacylglycerol + FA	Cytosol	Digestion of TG
Carnitine palmityl-transferase II	Acyl carnitine → Acyl CoA	Mitochondria	FA oxidation
Fatty acid cyclo-oxygenase	Arachidonic acid → Prostaglandin	Microsomes	Synthesis of prostaglandins

MCQ, Matching, True and False and Completion

Select and encircle the most appropriate answer or completion:

1. *Chylomicron remnants can be recognized by receptors present in liver cell membrane through:*
   A. Apolipoprotein E and apolipoprotein B_{48}
   B. Apolipoprotein E and apolipoprotein B_{100}.
   C. Apolipoprotein A-I.
   D. Apolipoprotein C-II.

2. *The plasma lipoprotein with the highest phospholipids content is:*
   A. Chylomicrons
   B. VLDL
   C. HDL
   D. LDL

3. *The plasma lipoprotein with the highest triacylglycerols content is:*
   A. Chylomicrons.
   B. VLDL.
   C. HDL.
   D. LDL.

4. *The plasma lipoprotein with the highest cholesterol content is:*
   A. Chylomicrons.
   B. VLDL.
   C. HDL.
   D. LDL.

5. *Ketone bodies are:*
   A. Succinyl CoA, α-ketoglutarate and acetyl CoA.
   B. Oxaloacetate, acetaldehyde and malate.
   C. Acetoacetate, β-hydroxybutyrate and acetone.
   D. Non of the above.

6. *An enzyme catalyzing the reaction: Lecithin + cholesterol → Cholesterol ester + Lysolethin is:*
   A. ACAT
   B. LCAT
   C. Lysolecithinase
   D. HMG CoA reductase

7. *Acetyl CoA is the precursor of the following compounds EXCEPT:*
   A. Carnitine.
   B. Cholesterol.
   C. Palmitate.
   D. Acetoacetate.

8. *Animal fed a high cholesterol diet shows decreased cholesterol synthesis by liver because of the inhibition of which one of the following enzyme?*
   A. HMG CoA synthetase
   B. HMG CoA reductase
   C. Mevalonate kinase
   D. HMG CoA lyase
   E. Sequalene synthetase

9. *The following hormones increase the mobilization of depot fat (lipolysis) EXCEPT:*
   A. Epinephrine and nor epinephrine..
   B. Insulin.
   C. Growth hormone.
   D. Glucagon.

10. *The immediate precursor of mevalonic acid is:*
   A. Mavalonyl CoA
   B. Mevalonyl pyrophosphate
   C. Acetoacetyl CoA
   D. β-Hydroxy, β-methylglutaryl CoA
   E. Acetyl CoA

11. *The net energy production in the form of ATP that can be produced by oxidation of one molecule of palmitate (16 C) is:*
   A. 12
   B. 36
   C. 129
   D. 146

12. *Succinyl CoA may undergo one of the following fate EXCEPT:*
   A. Heme synthesis
   B. Activation of ketone bodies
   C. Glycosaminoglycans synthesis.
   D. Gluconeogenesis

13. *NADPH+H' is important for the following reactions EXCEPT:*
   A. Respiratory burst
   B. Xenobiotics metabolism by cytochrome P450
   C. Reduction of pyruvate by lactate dehydrogenase
   D. Reduction of glutathione by glutathione reductase

14. *Apolipoprotein C-II activates:*
   A. LCAT
   B. ACAT
   C. Lipoprotein lipase.
   D. HMG-CoA reductase.

15. *Triacylglycerols present in plasma lipoproteins can be hydrolyzed by:*
   A. Lipoprotein lipase
   B. Hormone sensitive triacylglycerol lipase
   C. Heparin
   D. Pancreatic lipase
   E. Co-lipase

16. *Pancreatic lipase:*
   A. Hydrolyzes triacylglycerols to glycerol and fatty acids
   B. Is specific for the position 2 of triacylglycerols
   C. Is specific for the 1,3 ester linkage of triacylglycerols
   D. Specifically hydrolyzes cholesterol esters
   E. Is specific for phospholipids

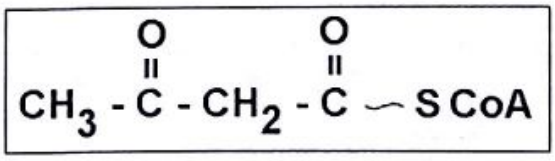
17. *A patient has a genetic defect of lipoprotein lipase. After eating high fat meal, one would expect to see plasma elevation of:*
   A. Chylomicrons
   B. VLDL
   C. LDL
   D. HDL
   E. Serum albumin associated lipid

18. *Which of the following is NOT a bile acid:*
   A. Glycocholic acid
   B. Deoxycholcholic acid
   C. Lithocholcholic acid
   D. Hippuric acid
   E. Glycochenodeoxycholic acid

19. *Which of the following changes would you expect in a patient with decreased activity of lipoprotein lipase?*
   A. Elevation of serum chylomicrons only.
   B. Elevation of both serum chylomicrons and VLDL.
   C. Elevation of HDL only.
   D. Elevation of serum LDL only.
   E. Elevation of both serum HDL and LDL.

20. *Cholesterol is the precursor of the following compounds EXCEPT:*
   A. Vitamin D_3.
   B. Bile acids.
   C. Testosterone.
   D. β-Hydroxy butyrate.
   E. Progesterone

21. *The compound shown beside is:*
   A. A bile salt
   B. Cholesterol
   C. A steroid hormone
   D. Vitamin D
   E. Acetoacetyl CoA



22. *In human, prostaglandins can be derived from:*
   A. Glucose
   B. Acetyl CoA
   C. Arachidonic acid
   D. Oleic acid
   E. Leukotrienes

23. *The site of ketogenesis is:*
   A. Lung
   B. Brain
   C. Liver
   D. Kidney

24. *The hormone that inhibits the mobilization of fat from adipose tissue is:*
   A. Epinephrine
   B. Norepinephrine
   C. Insulin
   D. Glucagon
   E. Growth hormone

25. *Activation of fatty acids requires:*
   A. ATP
   B. dATP
   C. GTP
   D. UTP
   E. CTP

26. *Acetyl CoA A carboxylase:*
   A. Requires pyridoxal phosphate for the carboxylation of acetyl CoA.
   B. Utilizes citrate as a substrate.
   C. Produces malonyl CoA, which is subsequently decarboxylated.
   D. Is located mainly in the mitochondrias.
   E. Is essentially key enzyme of fatty acid oxidation

27. *If one mole of $[CH_3\text{-}(CH_2)_{16}\text{-}COOH]$ is oxidized to CO_2 and H_2O in muscle mitochondria, the net number of moles of ATP produced will be?*
   A. *97*
   B. *114*
   C. *129*
   D. *146*
   E. *163*

28. *If one mole of palmitoleic (16:1 C) is oxidized to CO_2 and H_2O in muscle mitochondria, the net number of moles of ATP produced will be?*
   A. *97*
   B. *127*
   C. *129*
   D. *131*
   E. *146*

29. *How many moles of ATP are generated when one mole of β-hydroxybutyrate is oxidized to carbon dioxide and water in skeletal muscles?*
   A. *23*
   B. *24*
   C. *25*
   D. *26*
   E. *27*

30. *In the pathway leading to biosynthesis of acetoacetate from acetyl CoA in liver, which compound is the immediate precursor of acetoacetate?*
   A. β-Hydroxybutyrate
   B. Acetoacetyl CoA
   C. β-Hydroxybutyryl CoA
   D. Mevalonic acid
   E. β-Hydroxy-β-methylglutaryl CoA

31. *The key enzyme in the extramitochondrial synthesis of fatty acids is:*
   A. Condensing enzyme
   B. Hydratase
   C. Acetyl CoA carboxylase
   D. Acyl transferase
   E. Palmitoyl deacylase

32. *β-oxidation of fatty acids is dependant on the presence of all of the following enzymes EXCEPT:*
   A. Acyl CoA dehydrogenase
   B. Enoyl CoA hydratase
   C. β-Hydroxyacyl CoA dehydrogenase
   D. Acyl CoA acyltransferase
   E. Acetyl CoA carboxylase

33. *In the biosynthesis of fatty acids, which is the compound that transports the acetyl CoA out of the mitochondria into the cytosol?*
   A. Acetyl CoA itself
   B. Acetyl Carnitine
   C. Citrate
   D. Acetyl phosphate
   E. Acetyl choline

34. *The enzyme which catalyzes the activation of fatty acids, in the synthesis of triacylglycerols, is:*
   A. Condensing enzyme
   B. Acyl CoA synthetase
   C. Acyl CoA acyltransferase
   D. Lipase
   E. Glycerol kinase

35. *Ketosis results from:*
   A. Excessive production of propionyl CoA
   B. Excessive production of acetoacetyl CoA
   C. Excessive utilization of glucose
   D. Under utilization of lipids
   E. Hypercholesterolemia

36. *Complete oxidation of one molecule of stearic acid produces:*
   A. 20 ATP
   B. 96ATP
   C. 129 ATP
   D. 146 ATP

37. *β-Oxidation of stearic acid (18C) produces how many acetyl CoA molecule which later oxidized in citric acid cycle?:*
   A. 7 Acetyl CoA
   B. 8 Acetyl CoA
   C. 9 Acetyl CoA
   D. 12 Acetyl CoA

38. *All the following are sites of ketolysis EXCEPT:*
   A. Brain
   B. Liver
   C. Skeletal muscles
   D. Adipose tissue

39. *Succinyl CoA is required for:*
   A. Ketogenesis
   B. Heme synthesis
   C. Urea synthesis
   D. Cholesterol synthesis

40. *Carnitine palmitoyl transferase one is involved in fatty acid:*
   A. Synthesis
   B. Oxidation
   C. Activation
   D. Estrification
   E. Saponification

41. *A precursor for fatty acid synthesis is:*
   A. Propionyl CoA
   B. Malonyl CoA
   C. Acetoacetyl CoA
   D. Succinyl CoA

42. *All the following are lipotropic factors EXCEPT:*
   A. Essential fatty acids
   B. Folic acid
   C. Biotin
   D. Essential amino acids
   E. Choline

43. *The synthesis of palmitate from glucose in the liver:*
   A. Occurs in mitochondria.
   B. Utilizes $NADPH^+$ derived only from the pentose phosphate pathway.
   C. Is regulated mainly by isocitrate.
   D. Does not require biotin.
   E. Needs pantothenic acid that enters in the structure of acyl carrier protein.

44. *Which of the following sequences places the lipoproteins in the order of most dense to least dense (from left to right)?*
   A. HDL / VLDL / chylomicrons / LDL.
   B. HDL / LDL / VLDL / chylomicrons.
   C. LDL / chylomicrons / HDL / VLDL.
   D. VLDL / chylomicrons / LDL / HDL.
   E. LDL / chylomicrons / VLDL / HDL.

45. *During β-oxidation of even numbered fatty acid:*
   A. FAD is required to form a double bond.
   B. Carbon 2 of the fatty acid is oxidized to form a β-hydroxy compound.
   C. NAD^+ removes water from the β-hydroxy fatty acyl CoA intermediate.
   D. Two acetyl CoA molecules are produced in each turn of the β-oxidation.
   E. Propionyl CoA is the end product.

46. *If one mole of the compound (CH_3- $(CH_2)_{10}$ -COOH) is oxidized to CO_2 and H_2O in muscle mitochondria, the net number of moles of ATP produced will be:*
   A. 95
   B. 97
   C. 105
   D. 114
   E. 119

47. *The hepatic enzyme catalyzing the reaction: Fatty Acyl CoA + cholesterol → Cholesterol ester + CoA*
   A. LCAT
   B. ACAT
   C. Lysolecithinase
   D. HMG CoA reductase
   E. HMG CoA lyase

48. *In the synthesis of ketone bodies from fatty acids:*
   A. Carnitine transports the fatty acid across the plasma cell membrane.
   B. Acetoacetate and acetyl CoA are produced by cleavage of HMG CoA.
   C. The reactions occur in all tissues of the body.
   D. Activation of fatty acid is driven by the conversion of one ATP into ADP.
   E. Thiolase cleaves hydroxymethylglutaryl CoA (HMG CoA).

49. *In the course of the complete oxidation of acetoacetate:*
   A. Elevated insulin level is required.
   B. Activation by thiophorase enzyme is required.
   C. The reactions occur in the liver.
   D. A net of 30 ATP is produced.

50. *Phospholipids:*
   A. Always contain choline and glycerol.
   B. An important source of energy during fasting.
   C. Are a major component of membranes.
   D. Are not soluble in water.
   E. Are the main lipid conten of VLDL.

51. *In the palmitate synthesis its precursor is:*
   A. Mavalonyl CoA
   B. Acetoacetyl CoA
   C. Acetyl CoA
   D. β-Hydroxy, β-methylglutaryl CoA

52. *The product of extramitochondrial pathway for palmitate synthesis may undergo all the following EXCEPT:*
   A. May be converted into sphingosine base.
   B. May be converted into glucose.
   C. May be elongated to stearic acid.
   D. May be unsaturated to oleic acid.
   E. May be oxidized for energy production.

53. *β-oxidation of fatty acids involves a sequence of four reactions repeated several times. This sequence is best described by:*
   A. Oxidation - decarboxylation – dehydration – oxidation.
   B. Oxidation – dehydration – oxidation – thiolysis.
   C. Condensation – oxidation – dehydration – oxidation.
   D. Oxidation – hydration – oxidation – thiolysis.
   E. Condensation – hydration – oxidation – thiolysis.

54. *If one mole of the compound (CH_3- (CH_2) -(CH_2) -COOH) is oxidized to CO_2 and H_2O in muscle mitochondria, the net number of moles of ATP produced will be:*
   A. 27
   B. 37
   C. 95
   D. 129

55. *Which of the following changes would you expect in a patient with decreased activity of HMG CoA reductase enzyme?*
   A. Elevated serum cholesterol.
   B. Decreased serum cholesterol.
   C. Elevated serum triacylglycerols.
   D. Decreased serum triacylglycerols.

56. *An enzyme catalyzing the reaction: Fatty acyl CoA + cholesterol → Cholesterol ester + CoA is:*
   A. ACAT
   B. LCAT
   C. Lysolecithinase
   D. HMG CoA reductase

57. *Which of the following hormones inhibits lipolysis in adipose tissue:*
   A. Epinephrine and norepinephrin..
   B. Growth hormone.
   C. Glucagon.
   D. Insulin.

58. *In adiopose tissue:*
   A. An increase in intracellular cyclic AMP leads to increased release of triacylglycerol into the blood.
   B. Protein kinase is inactivated during fasting.
   C. Epinephrine stimulates the synthesis of triacylglycerols.
   D. Glucagon stimulates hormone sensitive triacylglycerols lipase.
   E. Glycerol kinase converts glycerol into glycerol-3-phosphate.

59. *The role of lipoprotein lipase is:*
   A. Digest dietary triacylglycerols in intestinal lumen
   B. Hydrolysis of triacylglycerols in adipose tissue
   C. Hydrolysis of triacylglycerols in plasma lipoproteins
   D. Hydrolysis of triacylglycerols in hepatocytes
   E. Activation of phospholipases.

60. *The rate controlling step in the synthesis of palmitate is catalyzed by:*
   A. Acetyl CoA carboxylase
   B. Fatty acid synthase
   C. HMG CoA synthase
   D. Hormone sensitive lipase
   E. Thiophorase

61. *All glycerol containing lipids are synthesized from:*
   A. Triacylglycerols
   B. Cephalin
   C. Phosphatidic acid
   D. Diacylglycerols
   E. Monoacylglycerols

62. *All the following statements are true about HDL EXCEPT:*
   A. HDL are involved in the transport of cholesterol from the periphery to the liver
   B. The major HDL apolipoproteins are AI and AII
   C. A high level of HDL –cholesterol is bad to one's health
   D. HDL appears antiatherogenic
   E. HDL is decreased after menopause

63. *The following are substrates for a carboxylase enzyme EXCEPT:*
   A. Pyruvate
   B. Acetyl CoA
   C. Propionyl CoA
   D. Acetoacetyl CoA

In the following questions indicate with clear (T) the true statements, and with clear (F) the false statements:

The following compounds are used to conjugate bile acids to form bile salts:

64. Glucuronic acid.
65. Glycine
66. Glutathione
67. Taurine
68. Acetyl CoA

Ketone bodies:

69. Can be used as a source of energy for muscles
70. Can be used as a source of energy for brain in starvation
71. Can be used as a source of energy for liver in starvation
72. If elevated in blood, it may cause metabolic alkalosis.
73. Are made during period of excessive lipolysis

The following compounds are intermediates in cholesterol biosynthesis:

74. Sequaline
75. HMG-CoA
76. Pregnanol
77. Lanosterol
78. Mevalonate

The sources of $NADPH+H^+$ are

79. *Pentose phosphate pathway*
80. *Uronic acid pathway for glucose oxidation*
81. *Action of cytosolic isocitrate dehydrogenase on isocitrate*
82. *Action of malic enzyme on malate.*
83. *Citric acid cycle*

A cyclooxygenase which is inhibited by aspirin is required for the production of:

84. Thromboxanes from arachidonic acid.
85. Leukotrienes from arachidonic acid.
86. Prostacyclin from arachidonic acid.
87. Arachidonic acid from Phospholipids.

Hypocholesterolemia is caused by:

88. Prolonged fasting
89. Hypothyroidism
90. Diabetes mellitus
91. Liver diseases
92. Diet rich in unsaturated fatty acids and poor in saturated fatty acids.

Ketone bodies:

93. Include only acetone and β-hydroxybutyrate
94. Can be used by liver cells as a source of energy
95. Are related to diabetes insipidus
96. May result from starvation
97. May be excreted in urine

The conversion of HMG CoA to mevalonic acid:

98. Requires $NADPH+H^+$
99. Is a key reaction in the synthesis of cholesterol
100. Is stimulated by insulin
101. Is a step in the synthesis of ketone bodies
102. May be regulated by both feed back inhibition and regulation

Excessive lipolysis is caused by:

103. Diabetes mellitus.
104. Diabetes insipidus
105. High carbohydrate diet
106. Starvation
107. Administration of nicotinamide and caffeine

In the synthesis of active vitamin D from 7-dehyrocholesterol:

108. The steroid ring structure is cleaved
109. Cholesterol is an intermediate compound
110. Ultraviolet light is required
111. Carbon 1 and 25 are hydroxylated in liver
112. Parathyroid hormone is required

Matching: For each set of numbered questions, choose the ONE BEST answer from the list of lettered options below it. An answer may be used once or more times, or not at all.

113. Synthesized from palmityl CoA and serine
114. Intermediate in ketone bodies synthesis
115. Product of odd number fatty acid oxidation
116. Dipalmityl lecithin

A. Lung surfactant
B. Propionyl CoA
C. Sphingosine
D. HMG-CoA

117. *Lecithin*
118. *Bile acid*
119. *Vitamin D_3*
120. *Carnitine*
   A. *β-Hydroxy γ trimethyl amino butyric acid*
   B. *Phosphatidyl choline*
   C. *Chenodeoxycholate*
   D. *1,25 Dihydroxycholecalciferol*

121. *Cholesterol synthesis*
122. *Cytoplasmic pathway for fatty acids synthesis*
123. *Lipolysis*
124. *Ketolysis*
125. *Thromboxanes synthesis*
   A. *HMG-CoA reductase.*
   B. *Acetyl CoA carboxylase*
   C. *Thiophorase*
   D. *Cyclooxygenase*
   E. *Hormone sensitive triacylglycerol lipase*

126. *Propionyl CoA carboxylase*
127. *Acyl CoA dehydrogenase*
128. *5- Lipoxygenase*
129. *Glucocerebrosidase*
   A. *Leukotrienes synthesis*
   B. *Gaucher's disease*
   C. *Oxidation of odd number fatty acids*
   D. *Fatty acid oxidation*

130. *Promotes mobilization of storage triacylglycerol*
131. *Helps in preventing fatty liver*
132. *Activates lipoprotein lipase*
133. *Is a source of methyl group for choline synthesis*
134. *Catalyzes cholesterol ester formation in liver*
   A. *Apolipoprotein CII*
   B. *ACAT enzyme*
   C. *Starvation*
   D. *Dietary choline*
   E. *S-Adenosyl methionine*

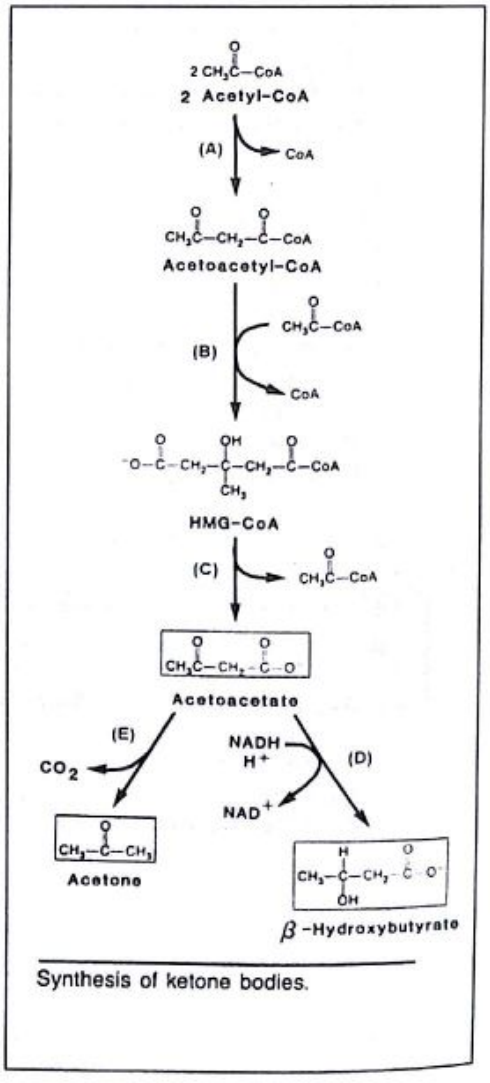
135. *Composed mainly of triacylglycerols*
136. *Composed mainly of cholesterol and cholesterol esters*
137. *Composed mainly of phospholipids and cholesterol*
138. *Eicosanoids*
139. *Acetoacetate, β-hydroxybutyrate and acetone*
   A. *Derived from arachidonic acid*
   B. *LDL*
   C. *Ketone bodies*
   D. *HDL*
   E. *Chylomicrons*

140. *Activator of LCAT enzyme*
141. *Binds with LDL receptors*
142. *Activator of lipoprotein lipase*
143. *Binds with chylomicrons receptors*
144. *Activator of pancreatic lipase*
   A. *Apo B $_{48}$*
   B. *Co-lipase*
   C. *Apo B $_{100}$*
   D. *Apo CII*
   E. *Apo A I*

145. *Gastric lipase*
146. *Pancreatic lipase*
147. *Lipoprotein lipase*
148. *Hormone sensitive triacylglycerol lipase*
149. *Acid lipase*
   A. *Degrades adipose tissue triacylglycerols.*
   B. *Degrades dietary triacylglycerol in infant.*
   C. *Acting on positions 1 and 3 of triacylglycerols.*
   D. *Acting on circulating triacylglycerols in blood.*
   E. *Removes fatty acids from lipids taken into cells during phagocytosis.*

150. *Hormone sensitive lipase*
151. *Lipoprotein lipase*
152. *Pancreatic lipase*
153. *Gastric lipase*
154. *Co-lipase*
   A. *Present in endothelial lining of some blood vessels*
   B. *Synthesized by pancreas*
   C. *Acts only in infant stomach*
   D. *Present in adipose tissue*
   E. *Activates pancreatic lipase*

In the synthesis of ketone bodies (figure beside):
155. *Spontaneous*
156. *HMG CoA synthase*
157. *3-hydroxybutyrate dehydrogenase*
158. *HMG CoA lyase*
159. *Thiolase*



Synthesis of ketone bodies.

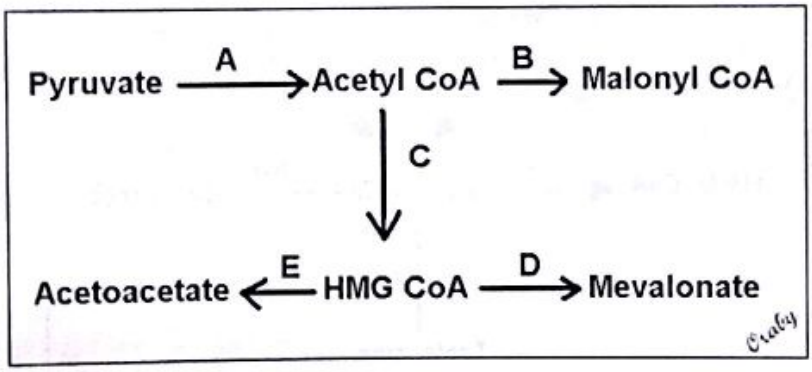
160. The lipid fraction increased in type I hyperlipoproteinemia is
161. The lipid fraction increased in type II hyperlipoproteinemia is
162. The lipid fraction increased in type III hyperlipoproteinemia is
163. The lipid fraction increased in type IV hyperlipoproteinemia is
164. The lipid fraction increased in type V hyperlipoproteinemia is

A. LDL
B. VLDL
C. Chylomicrons and VLDL
D. Chylomicrons and VLDL remnants

165. Contains 2% protein
166. Contains 10% protein
167. Contains 22% protein
168. Contains 50% protein
169. Contains 99% protein

A. HDL
B. VLDL
C. Chylomicrons
D. LDL
E. Fatty acid-albumin complex

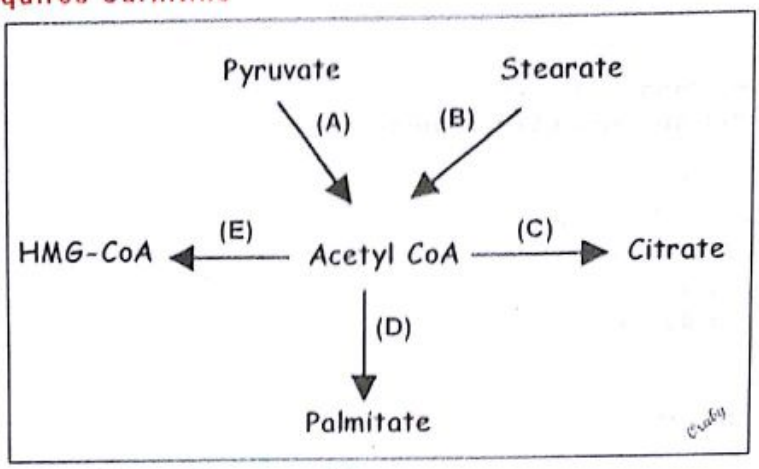
170. HMG CoA synthase
171. Requires TPP, Lipoic acid, CoASH, FAD and NAD+
172. Requires acetyl CoA carboxylase
173. Requires HMG CoA reductase
174. Requires HMG CoA lyase



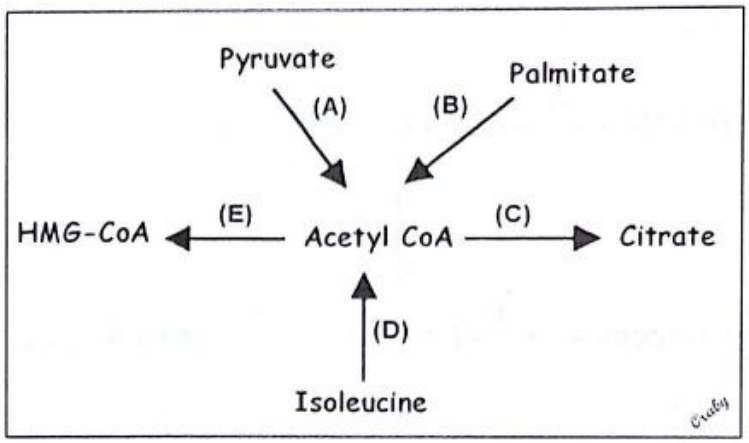
175. Chylomicrons
176. VLDL
177. LDL
178. HDL
179. Free fatty acids- Albumin complex

A. Synthesized in the kidney
B. Contain 99% protein
C. Contain apolipoproteins B_{48}
D. Pre β-Lipoproteins
E. Contain the good cholesterol
F. Derived from circulating IDL

180. Key enzyme is acetyl CoA carboxylase
181. 1st Reaction in Krebs' cycle
182. Requires TPP
183. Proceeds in cytosol to synthesize cholesterol
184. Requires Carnitine



185. Deficient in maple syrup disease
186. 1st Reaction in Krebs' cycle
187. Requires 5 coenzymes, all are vitamin B complex derivatives
188. Proceeds in hepatic mitochondria to synthesize acetoacetate
189. Requires Carnitine



Answer Key

MCQ:

1	2	3	4	5	6	7	8	9	10
A	C	A	D	C	B	A	B	B	D
11	12	13	14	15	16	17	18	19	20
C	C	C	C	A	C	A	D	B	D
21	22	23	24	25	26	27	28	29	30
E	C	C	C	A	C	D	B	B	E
31	32	33	34	35	36	37	38	39	40
C	E	C	B	B	D	C	B	B	B
41	42	43	44	45	46	47	48	49	50
B	C	E	B	A	A	B	B	B	C
51	52	53	54	55	56	57	58	59	60
C	B	D	A	B	A	D	D	C	A
61	62	63							
C	C	D							

True and false:

64	65	66	67	68	69	70	71	72	73
F	T	F	T	F	T	T	F	F	T
74	75	76	77	78	79	80	81	82	83
T	T	F	T	T	T	F	T	T	F
84	85	86	87	88	89	90	91	92	93
T	F	T	F	T	F	F	T	T	F

94	95	96	97	98	99	100	101	102	103
F	F	T	T	T	T	T	F	T	T
104	105	106	107	108	109	110	111	112	
F	F	T	T	T	F	T	F	T	

Matching:

113	114	115	116	117	118	119	120	121	122
C	D	B	A	B	C	D	A	A	B
123	124	125	126	127	128	129	130	131	132
E	C	D	C	D	A	B	C	D	A
133	134	135	136	137	138	139	140	141	142
E	B	E	B	D	A	C	E	C	D
143	144	145	146	147	148	149	150	151	152
A	B	B	C	D	A	E	D	A	B
153	154	155	156	157	158	159	160	161	162
C	E	E	B	D	C	A	C	A	D
163	164	165	166	167	168	169	170	171	172
B	C	C	B	D	A	E	C	A	B
173	174	175	176	177	178	179	180	181	182
D	E	C	D	F	E	B	D	C	A
183	184	185	186	187	188	189			
E	B	D	C	A	E	B			

Chapter 16 Protein Metabolism

1. *What is the enzyme mainly responsible for protein digestion in adult stomach?*
   A. Pepsin.
2. *What is the action of pepsin?*
   A. It hydrolyzes peptide bonds formed by carboxyl groups of aromatic amino acids as phenyl alanine, tyrosine and tryptophan.
3. *How pepsinogen is activated?*
   A. Through removal of N-terminal end of the enzyme by either (1) gastric HCl (2) pepsin itself.
4. *What are zymogens?*
   A. They are pro-enzymes, inactive at the time of secretion, but will be activated in the gastrointestinal tract.
5. *How the activation of zymogen is achieved?*
   A. By removing a small polypeptide part of the precursor molecule that masks the catalytic activity of the enzyme.
6. *What is its biological significance of producing enzymes as zymogens?*
   A. Zymogens prevent autodigestion of the cells.
7. *What are the important enzymes in pancreatic juice?*
   A. Trypsin, chymotrypsin, elastase, carboxypeptidase and collagenase.
8. *How trypsinogen is activated?*
   A. first by removal of a peptide from N-terminal end by enteropeptidase (produced by intestinal mucosa) then by trypsin itself.
9. *What is the action of trypsin?*
   A. It hydrolyzes peptide bonds formed by carboxyl groups of basic amino acids as arginine and lysine.
10. *How chymotrypsinogen is activated?*
    A. It is activated by trypsin.

11. *What are endopeptidases?*

A. Enzymes that act on peptide bonds inside the protein molecule, so that the protein becomes successively smaller and smaller units.

12. *Give some examples of endopeptidases.*

A. Trypsin and pepsin.

13. *What are exopeptidases?*

A. Enzymes that act at one end of the protein molecule, liberating amino acids sequentially, one at a time.

14. *Give examples of exopeptidases.*

A. Carboxypeptidase and aminopeptidase.

15. *What is carboxypeptidase?*

A. It is secreted by pancreas. It is a metalloenzyme containing zinc. It is an exopeptidase, spliting off carboxy terminal bond of the protein.

16. *What is aminopeptidase?*

A. It is an enzyme secreted by intestinal mucosal cells. It is an exopeptidase, spliting off amino terminal bond of the protein.

17. *What are mechanisms of amino acids absorption ?*

A. They include amino acid carrier proteins and glutathione.

18. *How many ATPs are utilized in amino acids absorption?*

A. For each amino acid, one ATP by using carrier proteins and 3 ATPs by using glutathione.

19. *What is γ - glutamyl transferase enzyme (GGT)?*

A. It catalyzes the transport of amino acids to inside the cell.

20. *What is the clinical significance of GGT?*

A. Its blood concentration increased in cholestasis (stop of bile flow) and chronic alcoholism.

21. *What are cathepsins?*

A. Intracellular proteases.

22. *In fasting state, nitrogen is transported from muscles as what form?*

A. In the fasting state, muscles release mainly alanine, glutamine and valine. They are taken up by liver (alanine), kidneys and intestine (glutamine) and brain (valine).

23. *What is transamination?*

A. It is transfer of α-amino group from an amino acid to α-keto acid to form the corresponding new amino acid and new α-keto acid.

24. *What are the physiological significance of transamination?*

A. (1) Synthesis of nonessential amino acids (2) Removal of amino group in the form of ammonia from most amino acids and glutamate through transdeamination.

25. *Transamination of oxaloacetic acid will give rise to what?*

A. Aspartic acid.

26. *Transamination reaction of pyruvate with glutamate results in the production of what substances?*

A. Alanine and α- ketoglutarate.

27. *What is the clinical significance of transamination?*

A. Blood transaminases are increased in liver and cardiac diseases.

28. *Give an example of transamination reaction.*

A. Glutamic acid + pyruvic acid ➔ α-keto glutarate + alanine.

29. *Transamination of glutamic acid produces what?*

A. α- ketoglutarate.

30. *What is the co-enzyme necessary for transamination reaction?*

A. Pyridoxal phosphate (PLP).

31. *What are the functions of pyridoxal phosphate in transamination?*

A. Pyridoxal phosphate (PLP) acts as an intermediate carrier of amino group to transfer it from amino acids to α-ketoglutarate.

32. *What is deamination?*

A. It is the removal of amino group from amino acids in the form of ammonia (NH3).

33. *Enumerate types of deamination:*

A. Oxidative deamination, non-oxidative deamination, and hydrolytic deamination.

34. *Which amino acid is oxidatively deaminated in liver?*

A. Glutamic acid.

35. *What is the reaction catalysed by L- glutamate dehydrogenase?*

A. Glutamate ➔ α- ketoglutarate + ammonia.

36. *What are the functions of L-glutamate dehydrogenase?*

A. Removal of NH_2 group of most amino acids as ammonia (transdeamination) and synthesis of non-essential amino acids.

37. *What is the co-enzyme required for L-glutamate dehydrogenase?*

A. NAD^+ or $NADP^+$.

38. *What is transdeamination?*

A. It is transamination of most amino acids with α-ketoglutarate to form glutamate. Then glutamate is deaminated to give ammonia (by L-glutamate dehydrogenase).

39. *What is the reaction catalysed by glutamine synthetase?*

A. Glutamate + ammonia ➔ glutamine. This reaction requires hydrolysis of ATP to ADP.

40. *What is the reaction catalysed by glutaminase?*

A. Glutamine ➔ glutamic acid + ammonia.

41. *Glutaminase enzyme is used for what function?*

A. Excretion of ammonia in kidney tubules.

42. *What is urea?*

A. It is the end product of protein and amino acid catabolism, where liver can convert toxic ammonia into nontoxic urea.

43. *What is the site of urea formation?*

A. Liver (in both cytosol and mitochondria).

44. *What is the key enzyme of urea synthesis?*

A. Carbamoyl phosphate synthase I.

45. *What are the two types of carbamoyl phosphate synthase (CPS)?*

A. CPS-I is involved in urea synthesis, CPS-II is required for pyrimidine synthesis. CPS-I is seen in mitochondria, while CPS-II is in cytosol.

46. *What is the normal plasma urea level?*

A. 20-50 mg/dl.

47. Plasma urea level is markedly increased in which condition?

A. Impairment of renal functions.

48. *What is the normal urinary excretion of urea?*

A. 20 - 40 grams/day.

49. *Nitrogen atoms in the urea are derived from what precursors?*

A. One from ammonia and another from aspartic acid.

50. *Give an account on relationship between tricarboxylic acid cycle (TCA) and urea cycle:*

A. CO_2 and ATP needed for urea formation are mostly produced in the course of TCA. Aspartate needed also for urea formation is derived from TCA from oxaloacetate by transamination. Fumarate produced in urea cycle can be oxidized in tricarboxylic acid cycle.

51. *Enumerate sources of ammonia:*

A. (1) By transdeamination of amino acids (liver), (2) by deamination of glutamine (kidneys), (3) by action of bacterial urease on urea present in intestinal fluids (intestine) and (4) catabolism of purines and pyrimidines.

52. *What are fate of ammonia:*

A. Formation of non-essential amino acids, glutamine and urea.

53. *Ammonia is immediately trapped in brain by what enzyme?*

A. Glutamine synthetase.

54. *What are causes of ammonia intoxication (hyperammonemia)?*

A. (1) Acquired hyperammonemia due to liver affection by cirrhosis or liver cell failure (2) Inherited hyperammonemia due to genetic deficiency of one of five enzymes of urea cycle.

55. *What are types of inherited ammonia intoxication?*

A. Hyperammonemia type I due to deficiency of **carbamoyl phosphate synthase I**, hyperammonemia type II due to deficiency of **ornithine transcarbamoylase**, citrullinemia due to deficiency of **arginosuccinic acid synthase**, argininosuccinate aciduria due to deficiency of **argininosuccinase** and argininemia due to deficiency of **arginase**.

56. *Explain why excess ammonia is toxic and may produce coma?*

A. At normal blood ammonia level, any ammonia reaches the brain is incorporated into glutamine formation by glutamine synthetase enzyme. In case of hyperammonemia, ammonia reacts not only with glutamate, but also with α-ketoglutarate by glutamate dehydrogenase enzyme. This depletes α-ketoglutarate, which is an essential intermediate of citric acid cycle. This results in a decrease ATP and energy production ➔ coma.

57. *What are amino acid forming oxaloacetate?*

A. Asparagine and aspartate.

58. *What are amino acid forming α-ketoglutarate?*

A. Glutamine, glutamate, proline, Arginine and histidine.

59. *What are amino acid forming pyruvate?*

A. Alanine, serine, glycine and cystine.

60. *What are amino acid forming formate?*

A. Phenylalanine and tyrosine.

61. *What are amino acid forming acetyl Co A and acetoacetyl CoA?*

A. Phenylalanine, tyrosine tryptophan, leucine, isoleucine, threonine, lysine, cycteine, cyctine, glycine, hydroxyproline and serine.

62. *What are amino acid forming succinyl CoA?*

A. Isoleucine, valine, threonine and methionine.

63. *Glycine is used for synthesis of what compounds?*

A. Serine, creatine, purines, heme, glutathione, bile salts, hippuric acid, collagen, neurotransmitter and glyoxylic acid.

64. *What is glutathione?*

A. γ- Glutamyl cysteinyl glycine.

65. *What are the functions of glutathione?*

A. Keeping RBC membrane integrity, carrying amino acids across membranes, detoxification of peroxides, activator for some enzymes and inactivation of insulin.

66. *What are amino acids forming glutathione?*

A. It is tripeptide formed of three amino acids, glutamate cysteine and glycine.

67. *What are enzymes involved in glutathione synthesis?*

A. Glutamyl cysteine synthetase and glutathione synthetase.

68. *What are purine atoms derived from glycine?*

A. C4, C5 and N7.

69. *For creatine synthesis, which amino acids are used?*

A. Glycine, arginine and methionine.

70. *Guanidoacetic acid is formed in which tissue and from what amino acids ?*

A. In kidney from arginine and + glycine.

71. *Where does methylation of guanidoacetic acid to form creatine occur?*

A. In liver.

72. *What are the functions of creatine?*

A. Creatine phosphate acts as a store of high energy phosphate in muscles and used as a source of energy during exercise.

73. *How creatinine is produced in the body?*

A. By spontaneous dehydration and dephosphorylation of creatine phosphate.

74. *Plasma creatinine level is markedly increased in which condition?*

A. In renal diseases and muscle disorders.

75. *What are the isoenzymes of creatine kinase (CK)?*

A. CK-MM (derived from muscles), CK-MB (derived from heart muscle) and CK-BB (derived from brain).

76. *What is the diagnostic importance of creatine kinase (CK) isoenzymes?*

A. CK-MM is elevated in muscle diseases (muscle atrophy), CK-MB is elevated in recent myocardial infarction, and CK-BB is elevated in damage of brain cells.

77. *What are the sources of oxalic acid in urine?*

A. Glycine and ascorbic acid.

78. *What are conditions of increased urinary excretion of oxalate?*

A. Primary hyperoxaluria and hypervitaminosis C.

79. *What are the metabolic errors of glycine in the body?*

A. Primary hyperoxaluria and glycinuria.

80. *What is primary hyperoxaluria?*

A. It is metabolic disease characterized by excessive excretion of oxalate unrelated to dietary intake of oxalate.

81. *What is the cause and effect of primary hyperoxaluria?*

A. It is due to failure of conversion of glyoxylate into formate. It may lead to excessive excretion of urinary oxalate and stone formation.

82. *What is the cause and effect of glycinuria?*

A. It is due to defect in renal tubular reabsorption of glycine. It leads to excessive glycine excretion with subsequent oxalate stone formation.

83. *What are the sources of glycine in the body?*

A. (1) **Serine** (by serine hydroxymethyl transferase enzyme), (2) **threonine** (by threonine aldolase enzymes), **and (3) glyoxylate** (by transamination).

84. *Which amino acids are both glucogenic and ketogenic?*

A. **Phenyl alanine, tyrosine, tryptophan, lysine and isoleucine.**

85. *Enumerate compounds given by tyrosine:*

A. **Catecholamines, melanin pigments, thyroid hormones. In intestine it produces putrefactive products as tyramine, phenol and cresol.**

86. *Where melanin is synthesized in the body?*

A. **In melanocytes present in eye (iris), skin and hair.**

87. *What is albinism?*

A. **It is a hereditary deficiency of tyrosine hydroxylase enzyme in melanocytes. This results in defective synthesis of melanin pigments in eye (iris) , skin and hair.**

88. *What is associated manifestation of ocular albinism?*

A. **Nystagmus (oscillatory ocular movement).**

89. *What is the immediate precursor of norepinephrine?*

A. **Dopamine.**

90. *In pheochromocytoma, urine contains what excess substance?*

A. **Vanillyl mandelic acid (VMA).**

91. *What is the catalytic activity of phenylalanine hydroxylase?*

A. **Conversion of phenylalanine to tyrosine.**

92. *Which one is essential, phenylalanine or tyrosine?*

A. **Phenylalanine is essential. Tyrosine is is non-essential, being synthesized in the body from phenylalanine.**

93. *When does tyrosine become essential?*

A. **If phenylalanine hydroxylase is deficient.**

94. *What are the co-enzymes required for phenylalanine hydroxylase?*

A. **Tetrahydro bioptrin and NADPH.**

95. *Which disease results from deficiency of phenylalanine hydroxylase?*

A. **Phenylketonuria.**

96. *What is the defective enzyme in phenylketonuria?*

A. **Phenylalanine hydroxylase.**

97. *What is atypical phenylketonuria?*

A. **It is due to deficiency of dihydrobiopterin reductase enzyme.**

98. *Phenylalanine, on transamination will give rise to what?*

A. **Phenylpyruvate.**

99. *What are the features of phenylketonuria?*

A. Mental retardation, hyperactivity, tremors, failure to walk and talk, failure to grow, IQ below 50, eczema and high blood phenyl alanine.

100. *What is the diagnostic importance of detection of neonatal blood phenylalanine?*

A. Early diagnosis of phenylketonuria before development of mental retardation.

101. *Phenylpyruvic acid is excreted in urine in which condition?*

A. Phenylketonuria.

102. *In phenylketonuria, urine will be positive for what test?*

A. Ferric chloride test.

103. *What is the catalytic activity of homogentisic acid oxidase?*

A. Transformation of homogentisic acid to maleylacetoacetate.

104. *Homogentisic acid is excreted in urine in which condition?*

A. Alkaptonuria.

105. *What is the defective enzyme in alkaptonuria?*

A. Homogentisic acid oxidase.

106. *Ochronosis is a manifestation of which condition?*

A. Alkaptonuria.

107. *A person's urine was found to turn black on standing and gave a positive* Benedict's test. He is likely to have what condition?

A. Alkaptonuria.

108. *What is the defect in tyrosinemia?*

A. Tyrosine-α-ketoglutarate transaminase and p-hydroxyphenyl-pyruvate oxidase.

109. *What are the important substances produced from tryptophan?*

A. Serotonin, melatonin, niacin, alanine, acetoacetate, indole and sketole.

110. *Tryptophan is deficient in which foodstuff?*

A. Maize and corn.

111. *Serotonin is derived from which amino acid?*

A. Tryptophan.

112. *Which amino acid will give rise to a vitamin?*

A. Tryptophan gives rise to niacin.

113. *What are the functions of niacin?*

A. Synthesis of NAD^+ and $NADP^+$ coenzymes that act as hydrogen carriers. Niacin also lowers plasma cholesterol.

114. *What is melatonin?*

A. Methyl acetyl serotonin.

115. *What is the importance of melatonin?*

A. It inhibits gonadal functions, has sleeping inducing effects, and inhibits synthesis of dopamine and GABA and regulation of circadian rhythm.

116. *What are characteristic features of malignant carcinoid syndrome (argentaffinoma)?*

A. Increased serotonin production on expense of niacin leading to niacin deficiency. This leads to manifestations of pellagra and hypertension.

117. *Pellagra is manifested in which conditions?*

A. Niacin deficiency, pyridoxal deficiency, tryptophan deficiency, Hartnup's disease and carcinoid syndrome.

118. *What is Hartnup's disease?*

A. It is a hereditary abnormality in tryptophan metabolism where the intestinal absorption and renal tubular reabsorption of this amino acid are impaired. This leads to tryptophan deficiency, and pellagra like symptoms → Mental retardation.

119. *Tryptophan is excreted in large quantities in which condition?*

A. Hartnup's disease.

120. *What is indican?*

A. Putrefaction product of tryptophan.

121. *What are the major functions of glutamic acid?*

A. Glutathione, glutamine, folic acid, γ-amino butyric acid, excitatory neurotransmitter, transamination, transdeamination, ammonia trapping in brain and activator for carbamoyl phosphate synthase I.

122. *Decarboxylation of glutamic acid will give rise to what?*

A. γ- Amino butyric acid (GABA).

123. *What is the function of γ- amino butyric acid (GABA)?*

A. It is an inhibitory neurotransmitter.

124. *Glutamic acid can be formed from which amino acids?*

A. Histidine, arginine, proline, glutamine.

125. *What are the functions of glutamine?*

A. Trapping ammonia in brain, regulation of acid base balance in kidney, synthesis of purines (nitrogen 3 and 9) and detoxification of phenylacetic acid.

126. *Is glutamine an amine?*

A. No, glutamine is an amide of glutamic acid.

127. *What is an amide?*

A. The extra carboxyl group (other than alpha carboxyl) can combine with ammonia to form the corresponding amide.

128. *α- Amino group of aspartic is incorporated into which compounds?*

A. Purines (1st nitrogen), pyrimidines (1st nitrogen and C4, 5, 6), and 1st nitrogen of urea.

129. *Which amino acids are required for both purine and pyrimidine synthesis?*

A. Aspartic acid and glutamine.

130. *What* are the *functions of aspartate?*

A. It enters in the formation of urea, purine (N_1), pyrimidine (N_1, C_4, C_5 and C_6), asparagine, neurotransmitter (excitatory) and β-Alanine.

131. *What is asparagine?*

A. is an amide of aspartic acid. It enters in the structure of some hormones like oxytocin and sources of ammonia.

132. *How asparagine is produced?*

A. Aspartic acid + ammonia (by asparagine synthetase and it requires ATP)).

133. *What is asparaginase?*

A. Enzyme catalysing the reaction asparagine to aspartic acid + ammonia.

134. *What is asparaginase function?*

A. Ammonia production in kidney tubules.

135. *What are the functions of histidine?*

A. Formation of histamine, carnosine, anserine and ergothionine.

136. *What is the decarboxylation product of histidine?*

A. Histamine.

137. *Which cells produce histamine?*

A. mast cells.

138. *Which is the vasodialator produced from histidine?*

A. Histamine.

139. *What is the significance of histamine?*

A. Vasodilatation, contraction of smooth muscles of bronchi, stimulation of gastric secretions. Histamine also acts as neurotransmitter.

140. *What is the clinical significance of histamine?*

A. It is a powerful vasodilator and mediator of allergic reactions (anaphylaxis).

141. *What is figlu?*

A. Formimino glutamic acid, it is a product of histidine metabolism.

142. *What is Figlu excretion test?*

A. In folic acid deficiency, there is a block in histidine metabolism, and figlu is excreted in large quantities in urine.

143. *Tetrahydro folic acid is used for what purpose?*

A. It is the carrier of one carbon fragments.

144. *Name one carbon compounds.*

A. Formyl, formimino, methenyl, methyl, methylene, and methyl groups.

145. *Which amino acids are the donors to one carbon pool?*

A. Serine, glycine, tryptophan, histidine and choline.

146. *One carbon units are used for synthesis of what?*

A. C2 and C8 of purine, serine, dTMP, choline, creatine, epinephrine.

147. *Name some monoamines.*

A. Histamine, dopamine and serotonin.

148. *What is histinemia?*

A. This is a hereditary disease due to deficiency of histidinase enzyme. It is characterized by mental retardation and speech defects.

149. *What are the important substances produced from serine?*

A. Phosphoproteins, sphingosine base, cysteine, purines, glycine, ethanolamine, choline, phosphatidyl serine and pyruvate.

150. *On decarboxylation, serine will produce what?*

A. Ethanolamine and choline.

151. *What is the product of transamination of alanine?*

A. Pyruvic acid.

152. *What is the significance of glucose-alanine cycle?*

A. During starvation, alanine is released from muscle, and is taken up by liver. In liver alanine is transaminated to pyruvate, and pyruvate undergoes gluconeogenesis to give glucose.

153. *How alanine amino acid is related to carbohydrate metabolism?*

A. Through glucose-alanine cycle.

154. *Which amino acid has two optically active (asymmetric) carbon atom?*

A. Threonine.

155. *Lysine is deficient in which foodstuffs?*

A. Cereals.

156. *What are functions of aspartate?*

A. Synthesis of urea, purine (N_1), pyrimidine (N_1, C_4, C_5 and C_6), asparagine, neurotransmitter (excitatory) and β-Alanine.

157. *Carnitine is synthesized from which amino acid?*

A. Lysine.

158. *Nitric oxide synthase system needs which coenzymes?*

A. FAD, FMN , tetrahydro biopterine, NADPH.

159. *What are the functions of arginine in the body?*

A. Formation of urea, creatine, nitric oxide (NO) and arginine phosphate (arginine P).

160. *What arginine phosphate?*

A. It is present in muscles and acts as a source of energy in animals (invertebrates).

161. *What are the major functions of nitric oxide?*

A. Vasodilator, smooth muscle relaxant and neurotransmitter.

162. *What is the precursor of nitric oxide?*

A. Arginine.

163. *Which amino acid will give rise to polyamines?*

A. Ornithine.

164. *What are polyamines?*

A. Polyamines are putrescine, spermine and spermidine.

165. *What are the functions of ornithin?*

A. Formation of urea, spermidine and spermine. It is used also in detoxication.

166. *What are the functions of polyamines?*

A. They are growth factors; their concentration is increased in cancer.

167. *Valine enters in which metabolic pathway?*

A. Valine is glucogenic.

168. *Leucine enters in which metabolic pathway?*

A. Leucine is ketogenic.

169. *Isoleucine joins in which metabolic pathway?*

A. Isoleucine is partly glucogenic and partly ketogenic.

170. *Branched chain ketoacids are excreted in urine in what condition?*

A. Maple syrup urine disease.

171. *What is the defect in maple syrup urine disease?*

A. Deficient oxidative decarboxylation of branched chain keto acids.

172. *Which is the purely ketogenic amino acid?*

A. Leucine.

173. *What are important aminoacidurias, which cause mental retardation?*

A. Phenyl ketonuria, homocystinuria, maple syrup urine disease.

174. *What are the functions of proline?*

A. Formation of hydroxyproline and collagen synthesis.

175. *What are the functions of β-alanine?*

A. Formation of pantothenic acid, anserine, carnosine and coenzyme A.

176. *What are the functions of thioethylamine?*

A. Formation of coenzyme A and acyl carrier protein.

177. *What are the functions of threonine?*

A. Formation of phosphoproteins and glycine.

178. *What are the functions of cysteine?*

A. Formation of glutathione, taurine, thioethylamine, cysteine, keratins, active center of many enzymes and detoxication.

179. *What are the functions of taurine?*

A. Taurine combines with sodium cholic acid to form sodium taurocholic acid. It is one of bile salts, which are important for digestion and absorption of lipids.

180. *What are the functions of methionine?*

A. Formation of S-adenosyl methionine, cysteine, spermidine, spermine. Methionine acts as lipotropic factor.

181. *What is transmethylation?*

A. Transmethylation is the transfer of methyl group from methyl donor to methyl acceptor.

182. *Give example of methyl donor?*

A. Active methionine (S-adenosyl methionine) and active choline (betaine).

183. *What is the methyl donor in transmethylation reaction?*

A. S-adenosyl methionine.

184. *What are important substrates for transmethylation reactions (methyl acceptors)?*

A. Homocysteine (to give methionine), uracil (to give thymine), norepinephrine (to give epinephrine), guanidoacetic acid (to give creatine), and carnosine (to give anserine).

185. *What is the defective enzyme in homocystinuria?*

A. Cystathionine β-synthase enzyme.

186. *What are the characteristic features of homocystinuria?*

A. Mental retardation, osteoporosis, dislocation of eye lens and thromboses.

187. *Homocystinuria is due to abnormal metabolism of which amino acid?*

A. Methionine.

188. *What is cystathionuria?*

A. This is accumulation and excretion of cystathionine due to deficiency of cystathionase enzyme. No clinical symptoms are present.

189. *What are amino acids used in Detoxication?*

A. Glycine, cysteine and glutamic acid.

190. *What are amino acids used in synthesis of vitamins?*

A. Tryptophan gives nicotinic acid, β-alanine gives pantothenic acid, glutamic acid gives folic acid and serine gives choline.

191. *What are amino acids used in synthesis of hormones?*

A. Tyrosine gives catecholamines and thyroxin and tryptophan gives melatonin.

192. *What is nitrogen balance?*

A. Nitrogen intake (in the form of dietary proteins) is equal to nitrogen loss from the body (in urine, feces and in milk).

193. *What is positive nitrogen balance?*

A. Nitrogen intake is greater than nitrogen loss. It occurs where the formation of tissue proteins is increased e.g. growing children and muscle training.

194. *What is negative nitrogen balance?*

A. Nitrogen intake is less than nitrogen loss, It occurs where the breakdown of tissue proteins is increased e.g. diabetes mellitus and starvation.

195. *What is hypoproteinosis?*

A. It is a condition of dietary proteins deficiency.

196. *What are causes of hypoproteinosis?*

A. In adult hypoproteinemia and In infants kwashiorkor and marsmus.

197. *What is kwashiorkor?*

A. A nutritional disease resulting from deficiency of only dietary protein children aged 1-3 years. It leads to growth retardation, anemia, vomiting and anorexia (loss of appetite).

198. *What is marsmus?*

A. This is a disease resulting from deficiency of dietary protein together with dietary carbohydrate and fat.

199. *What is Alzheimer?*

A. It is a neurological disorder resulting from deposition a protein known as β-amyloid. It is insoluble protein. It leads to dementia.

200. *What is a prion disease?*

It is also called spongiform encephalopathies (or mad cow disease). It is a neuro degenerative disease due to aggregation and deposition of *infective* protein known as *prion protein*. It is insoluble protein.

201. *Which amino acids act as an inhibitory neurotransmitter?*

A. Glutamic acid and glycine.

202. *What is meant by neurotransmitter?*

A. Neurotransmitter is a chemical substance synthesized and released by one neuron (by presynaptic terminal) into a specific receptor or an adjacent cell (post synaptic).

203. What is chemical nature of neurotransmitter?

A. Amino acids (glycine, glutamate and aspartate), amino acid derivatives (catecholamines, serotonin, GABA and histidine) or peptides (substance P & endogenous opioid peptides).

204. What are neuropeptides?

A. They include endogenous opioids, substance P, somatostatin, thyrotropin releasing hormone (TRH), corticotropin releasing hormone, vasoactive intestinal polypeptide, cholecystokinin and neurotensin.

205. What are endogenous opioids?

A. They include endorphins, enkephalins, dynorphins), They are large peptides present mainly in the pituitary and hypothalamus.

206. What is function of β-endorphin?

A. β-Endorphins are 18-30 times more potent than morphine. They inhibit the release of substance P i.e. relieve pain.

207. What is Parkinson's disease?

A. It is due to decrease of dopamine neurotransmitter in mid brain ➔ *(gradual degeneration of certain cerebral area of substania nigra present in mid brain)*. It occurs over 60 years. It includes static tremors (pills rolling). These tremors interfere with motor functions of skeletal muscles.

208. Succinyl CoA is formed from which substrates?

A. Isoleucine, valine, methionine, and odd chain fatty acids.

209. Fumarate is produced from which substances?

A. A. Arginino succinate. B. Phenyl alanine.

210. Alpha keto glutaric acid is formed from which substances?

A. Glutamic acid, histidine, arginine, and proline.

211. Aspartic acid enters the TCA cycle at which level?

A. Oxaloacetate.

212. Compare between Carbamoyl phosphate synthetase(CPS) I & II:

	CPS I	CPS II.
Function	Urea synthesis	Pyrimidine synthesis
Site	Mitochondria of liver cells	Cytosol of most tissues
Substrates	Ammonia, CO_2 and ATP	Glutamine,CO_2 and ATP

213. Compare between triiodothyronine (T3) and reverse triiodothyronine (rT3):

	T3	rT3
Structure	3, 5 ,5' Triiodothyronine	3,3',5' Triiodothyronine
Bilogical activity	Active	Inactive

214. Compare between L-amino acid oxidase and L-glutamate dehydrogenase.

	L-Amino acid oxidase	L-glutamate DH
Site	Liver and kidney	Cytosol and mitochondria of most tissues
Substrate	L-Amino acids	L-Glutamate
Coenzyme	FMN	NAD^+ or $NADP^+$
H_2O_2	Produced	Not produced
Reaction	Irreversible	Reversible
Type of dehydrogenase	Aerobic	Anaerobic
Cytochrome system.	Needed	Not needed

MCQ, Matching, True and False and Completion

Select and encircle the most appropriate answer or completion:

1. *Adrenaline and nor-adrenaline are formed from:*
   A. Epinephrine
   B. Tyrosine
   C. Cysteine
   D. Tryptophan

2. *Thyroxin is derived from:*
   A. Tyrosine
   B. Tryptophan
   C. Taurine
   D. Tryptamine
   E. Methionine

3. *Tyrosine is:*
   A. An essential amino acid
   B. A precursor in the biosynthesis of serotonin
   C. A precursor in the biosynthesis of γ-aminobutyric acid (GABA)
   D. Both ketogenic and glucogenic
   E. A precursor in the biosynthesis of thiamin

4. *Transaminases:*
   A. Usually require glutamine as one of the reacting substrates.
   B. Catalyze reactions that result in the production of ammonia.
   C. Catalyze irreversible reactions.
   D. Require pyridoxal phosphate as coenzyme.
   E. Are unable to catalyze transamination reactions with essential amino acids.

5. *All the following about transaminases are true EXCEPT:*
   A. Direct produce free ammonia
   B. Need pyridoxal phosphate as a coenzyme
   C. Catalyze reversible reactions.
   D. Function to synthesize nonessential amino acids
   E. Used for diagnosis of some diseases

6. *Phenylalanine and tyrosine enter the citric acid cycle after degradation to:*
   A. Pyruvate
   B. Fumarate
   C. Succinyl CoA
   D. α-Ketoglutarate
   E. Citrate

7. *S-Adenosylmethionine is a methylating agent that transfers a methyl group to:*
   A. Acetate
   B. Homocysteine
   C. Norepinephrine
   D. Pyruvic acid
   E. Testosterone

8. *Serine is converted to ethanolamine by the removal of:*
   A. Hydrogen
   B. Oxygen
   C. Carbon dioxide
   D. Ammonia
   E. A carboxyl group

9. *Gamma decarboxylation of aspartic acid produces:*
   A. Asparagine
   B. β-Alanine
   C. γ-Amino butyric acid
   D. Glutamic acid
   E. Serine

10. *Tryptophan is NOT used in the biosynthesis of:*
   A. Niacin
   B. Serotinine
   C. Melatonine
   D. Norepinephrine
   E. Indol

11. *Glycine is needed for the formation of:*
   A. Serotonin
   B. Glutathione
   C. Pyrimidine bases
   D. Thyroxine
   E. Melatonin

12. *Aspartic acid is the source of:*
   A. One nitrogen atom of urea
   B. The two nitrogen atoms of urea
   C. The carbon atom of the urea
   D. The oxygen atom of the urea

13. *If a normal adult is placed on a diet deficient only in phenyalanine, which of the following statement is CORRECT:*
   A. Signs and symptoms of phenylketonuria will appear.
   B. Signs and symptoms of myxedema will appear.
   C. Tyrosine in the diet cannot be used to synthesize catecholamine.
   D. Tyrosine becomes an essential amino acid.

14. *In relation to proteins, which of the following statements is CORRECT:*
   A. The end product of all amino acids catabolism is urea.
   B. All essential amino acids are glycogenic.
   C. Ornithine, citrulline and β-alanine are found in tissue proteins.
   D. In the presence of adequate dietary sources of tyrosine, phenylalanine is not an essential amino acid.

15. *Amino acids considered non-essential for human are:*
   A. Those incorporated into protein.
   B. Those not synthesized in the body.
   C. Those synthesized post-translationally.
   D. Those cannot be transaminated.
   E. Those cannot be deaminated.

16. *In the formation of urea, all the followings are correct EXCEPT:*
   A. Aspartate supplies one of the nitrogen found in urea.
   B. Fumarate is produced.
   C. Genetic deficiency of any enzyme can lead to hyperammonemia.
   D. Carbamoyl phosphate synthase I requires N-acetyl glutamate as allosteric effector.
   E. The site of the urea formation is the kidneys.

17. *All of the followings are correct about ornithine EXCEPT:*
   A. Plays a major role in urea cycle.
   B. Is a precursor of spermin and spermidine.
   C. Can be used in detoxication of phenyl acetyl CoA.
   D. Is a source of N1 of purine bases.
   E. It is non protein amino acid.

18. *Removal of amino groups from amino acids in man is performed mainly by:*
   A. Transamination
   B. Oxidative deamination
   C. Transdeamination
   D. Non-oxidative deamination
   E. Hydrolytic deamination

19. *Serine is important for synthesis of all of the following EXCEPT:*
   A. Phosphoproteins
   B. Sphingosine
   C. Carnitine.
   D. Choline.
   E. Glycine

20. *All of the followings are true about the branched chain amino acids EXCEPT:*
   A. All are essential amino acids.
   B. All are glucogenic amino acids.
   C. Are metabolized initially in brain, muscles and adipose tissues.
   D. Metabolic error leads to Maple syrup urine disease.
   E. Are catabolized into succinyl CoA, acetyl CoA and acetoacetyl CoA.

21. *Which of the following amino acids can be metabolized to fatty acids in mammals:*
   A. Leucine
   B. Glycine
   C. Methionine
   D. Arginine
   E. Histidine

22. *Which of the following amino acids is synthesized only posttranslationaly, after incorporation of a precursor into a polypeptide?*
   A. Proline
   B. Hydroxyproline
   C. Glutamate
   D. Serine
   E. Glycine

23. Which amino acid undergoes transamination to form oxaloacetate:
A. Alanine
B. Glycine
C. Aspartate
D. Lysine
E. Ornithine

24. Which amino acid undergoes transamination to form glyoxylic acid:
A. Alanine
B. Glycine
C. Glutamate
D. Aspartate
E. Lysine

25. All the followings about the glutamate dehydrogenase are true EXCEPT:
A. It catalyzes a reversible reaction
B. Its coenzyme is FAD.
C. It catalyzes the synthesis of non-essential amino acids.
D. It catalyzes the removal of amino group in the form of ammonia.
E. It is allosterically inhibited by ATP, GTP and $NADH+H^+$.

26. Alkaptonuria is due to the absence of:
A. Homogentisic acid oxidase
B. Tyrosine oxidase
C. Tyrosine hydroxylase
D. Phenylalanine hydroxylase
E. Dopa decarboxylase

27. All the following about phenylketonuria are true EXCEPT:
A. Is associated with mental retardation
B. Is due to deficiency of phenylalanine in diet
C. Is due to inherited deficiency of phenylalanine hydroxylase
D. May result from deficiency of dihydrobiopterin reductase
E. Is associated with excretion of phenylpyruvate and phenyllactate

28. All the following about tyrosinemia are true EXCEPT:
A. Is associated with liver cirrhosis or failure
B. Is due to inherited deficiency of tyrosine α-ketoglutarate transaminase
C. May result from deficiency of parahydroxy phenylpyruvate oxidase
D. Has two forms acute and chronic
E. Prevented by feeding infant with diet low in phenylalanine.

29. The metabolism of tryptophan results in production of a substance that:
A. Forms heme.
B. Prevents albinism.
C. Causes vasoconstriction.
D. Causes vasodilatation
E. Causes phenylketonuria.

30. An essential amino acid in man is:
A. L-proline
B. L-tyrosine
C. L-serine
D. L-methionine
E. L-glutamate

31. *Albinism is due to the absence of:*

A. Tyrosine hydroxylase
B. Dopa oxidase
C. Tryptophan pyrrolase
D. Homogentisic acid oxidase.
E. parahydroxy phenylpyruvate oxidase

32. *All the following statements concerning the degradation of amino acids are true EXCEPT:*

A. Leucine leads to the production of succinyl CoA, via propionyl CoA.
B. Histidine leads to the production of acetyl CoA via glycine and acetate.
C. Phenyl alanine leads to the production of fumarate via tyrosine and homogentisic acid.
D. Aspartate lead to the production of oxaloacetate via α-ketoglutarate.
E. Tryptophan leads to the production of 5-hydroxy indolacetic acid via serotonin.

33. *Which of the following proteolytic enzymes has the greatest effect on the activity of other proteolytic enzymes involved in protein digestion?*

A. Trypsin
B. Chymotrypsin
C. Carboxypeptidase
D. Elastase
E. Aminopeptidase

34. *Which of the following statements about the synthesis of carbamoyl phosphate by carbamoyl phosphate I is INCORRECT?*

A. The reaction is allosterically activated by N-acetyl glutamate.
B. The reaction requires hydrolysis of two high energy phosphate bonds.
C. The reaction is considered the rate limiting step in the urea cycle.
D. The reaction is reversible.
E. The reaction occurs in the mitochondria of liver cells.

35. *Which of the followings is synthesized from an essential amino acid?*

A. Alanine
B. Glutamate
C. Proline
D. Tyrosine
E. Aspartate

36. *Concerning glutamine, which one of the following statements is INCORRECT?*

A. Its synthesis needs glutamine synthetase.
B. It trapped ammonia in a nontoxic form in the brain.
C. It is one reactant of all transamination reaction.
D. It is the source of N_3 and N_9 of purine bases.
E. It may be used in regulation of acid base balance in the kidney.

37. *Aspartic acid is incorporated in the synthesis of:*

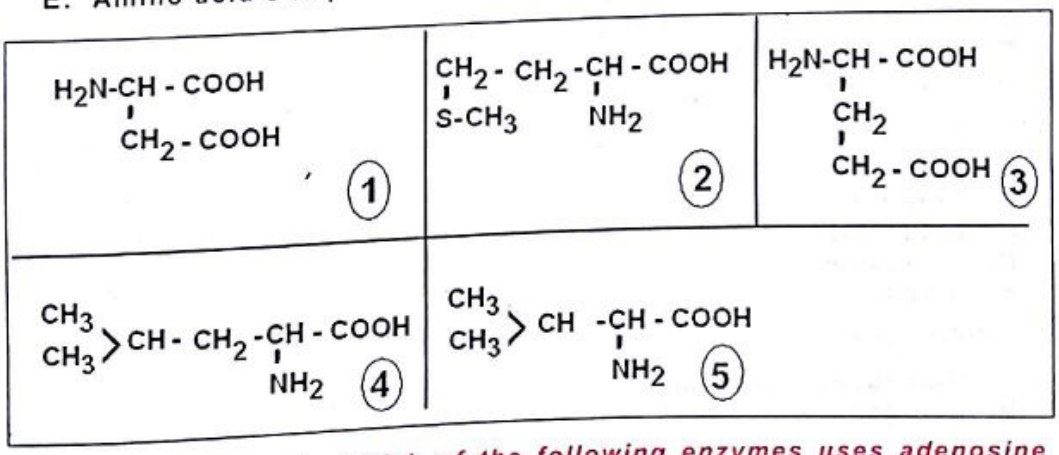
A. Porphyrins
B. Steroids
C. Sphingolipids
D. Pyrimidines
E. Coenzyme A

38. *Which one of the following statements concerning urea cycle is CORRECT?*
   A. It occurs in the cytosol of intestinal cells.
   B. It utilizes 2 ATP molecules.
   C. N-acetylaspartate is an activator of carbamoyl phosphate synthase I.
   D. The immediate precursor of urea is citrulline.
   E. In patients with renal failure, urea levels are elevated.

39. *Which one of the following statements concerning protein digestion is CORRECT?*
   A. Chymotrypsin acts on peptide bonds formed by the amino acid glycine.
   B. Trypsin is an exopeptidase that liberates free amino acids.
   C. Pepsinogen is activated by enteropeptidase.
   D. Pepsin is an endopeptidase secreted by the pancreas.
   E. Trypsin can act as an activator for all zymogens of pancreatic proteases.

40. *Which one of the following statements is CORRECT?*
   A. In human, the end product of amino acids catabolism is uric acid.
   B. Elastase is the enzyme that is acting on elastine.
   C. Rennin acts on casein in presence of calcium ions to form milk clot.
   D. In fasting state, plasma amino acids are derived mostly from liver.
   E. Transaminase using alanine and α-ketoglutarate as substrates produces glutamate and oxaloacetate.

41. In the figure below, which one of the following statements is CORRECT?
   A. Amino acids 1 and 2 are glucogenic.
   B. Amino acids 2, 3 and 4 are essential amino acids.
   C. Amino acids 4 is glucogenic.
   D. Amino acid 3 is catabolized to pyruvate after transamination.
   E. Amino acid 5 is pure ketogenic.



42. *In urea synthesis, which of the following enzymes uses adenosine triphosphate (ATP)?*
   A. Arginase
   B. Ornithin transcarbamoylase
   C. Arginosuccinate synthase
   D. Arginosuccinase
   E. Fumarase

43. *Which of the following amino acids is formed by transamination of a member of the glycolytic pathway:*

A. Methionine
B. Lysine
C. Serine
D. Tyrosine
E. Valine

44. *Among 100 grams free amino acids distributed throughout the body, the amino acids found in the highest concentration are:*

A. Alanine and β-alanine
B. Glutamate and glutamine
C. Alanine and glycine
D. Phenylalanine and tyrosine
E. Aspartate and asparagines

45. *The decarboxylation of which following amino acids produces a vasodilating compound?*

A. Arginine
B. Aspartic acid
C. Histidine
D. Glutamine
E. Proline

46. *In the urea cycle:*

A. Carbamoylphosphate is derived directly from glutamine and CO_2
B. Ornithin directly reacts with aspartate to generate arginosuccinate
C. Ornithine directly reacts with carbamoylphosphate to form citrulline
D. The α-amino group of arginine forms one of the nitrogens of urea
E. N-Aetylglutamate is a positive allosteric effector of ornithine transcarbamoylase

47. *Conversion of creatine into creatinine is:*

A. An enzymatic dehydration reaction
B. Non-enzymatic dehydration reaction
C. Reversible reaction
D. Hydrolytic reaction

48. *Tryptophan is the precursor of:*

A. Melanin
B. Creatine
C. Epinephrine
D. Thyroxine
E. Niacin

49. *Histidine is:*

A. Glucogenic amino acid
B. Ketogenic amino acid
C. Glucogenic and ketogenic amino acid
D. Non essential amino acid
E. Imino acid

50. *Epinephrine is formed from norepinephrine by:*

A. Methylation
B. Hydroxylation
C. Carboxylation
D. Oxidative decarboxylation
E. Oxidative deamination

51. *Phenylketonuria is a genetic defect caused by the absence of the enzyme:*
   A. A Ketoacid decarboxylase
   B. Tyrosinse
   C. Phenylalanine hydroxylase
   D. Homogentisic acid oxidase
   E. Alanine transaminase

52. *Which of the following statements is INCORRECT:*
   A. All essential amino acids are glycogenic
   B. In deficiency of phenyl alanine hydroxylase, tyrosine becomes essential
   C. The end product of all amino acids catabolism is urea
   D. Pyridoxal phosphate is the coenzyme for transamination
   E. Reverse T_3 is biologically inactive hormone

53. *Hippuric acid is a detoxication product of:*
   A. Aspirin
   B. Benzoic acid
   C. Bromobenzine
   D. Testosterone
   E. Phenyl acetic acid

54. *Hartnup's disease is an inborn error in metabolism of the amino acid:*
   A. Histidine
   B. Arginine
   C. Tryptophan
   D. Isoleucine
   E. Tyrosine

55. *Melatonine is derived from:*
   A. Tyrosine
   B. Phenyl alanine
   C. Tryptophan
   D. Taurine
   E. Tryptamine

56. *In mammals, tryptophan is a precursor of all of the following EXCEPT:*
   A. Nicotinate
   B. Serotonin
   C. Histamine
   D. Melatonin
   E. Niacin

57. *The metabolism of tryptophan results in production of a substance that:*
   A. Acts as a neurotransmitters
   B. Forms heme
   C. Prevents albinism
   D. Causes tyrosinosis
   E. Causes vasodilatation

58. *All the followings are true about tryptophan EXCEPT:*
   A. An essential amino acid
   B. Is ketogenic and glucogenic
   C. A precursor in the biosynthesis of serotonin
   D. A precursor in the biosynthesis of melatonin
   E. A precursor in the biosynthesis of melanin

59. *All the followings occur in humans EXCEPT:*
   A. Serine → Cysteine
   B. Homocysteine → Methionine
   C. Phenylalanine → Tyrosine
   D. Arginine → Arginine phosphate
   E. Proline → Glutamate

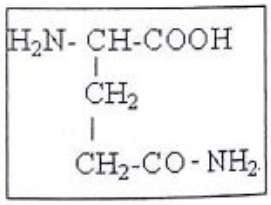
60. *Which of the following is NOT involved in the biosynthesis of creatine phosphate?*
   A. Pyridoxal phosphate
   B. ATP
   C. Methionine
   D. Glycine
   E. Arginine

61. *Which enzymatic step is NOT involved in the biosynthesis of epinephrine:*
   A. A methylation
   B. An aliphatic hydroxylation
   C. An aromatic hydroxylation
   D. A carboxylation
   E. A decarboxylation

62. *The major amino acid that is released from muscle and converted to glucose in the liver is:*
   A. Alanine
   B. Glutamine
   C. Valine
   D. Aspartate
   E. Glutamate

63. *Each of the following statements about the kidney is correct EXCEPT:*
   A. It uses ammonia released from glutamine to buffer acids
   B. It converts glutamine to glutamate and free amonnia
   C. During fasting, it provides plasma alanine and serine
   D. It synthesizes most of the urea that is excreted in urine
   E. It contains one enzyme responsible for creatine synthesis

64. *The structure beside:*
   A. Is a dipeptide
   B. Is glutamine
   C. Is given the abbreviation GABA
   D. Can involved in linking phosphate to a protein chain
   E. Is essential dietary component for human



65. *Which of the following 2 vitamins are involved in transformation of serine to glucose:*
   A. B_{12} and nicotinamide
   B. Folic acid and Pyridoxal phosphate
   C. Folic acid and B_{12}
   D. Riboflavin and niacin
   E. Pyridoxal phosphate and B_{12}

66. *The enzyme enteropeptidase is important in the intestinal digestion of dietary protein because it converts:*

A. Pepsinogen to pepsin
B. Procarboxypeptidase A to carboxypeptidase A
C. Trypsinogen to trypsin
D. Procarboxypeptidase B to carboxypeptidase B
E. Chemotrypsinogen to trypsinogen.

67. *The principle nitrogenous urinary excretion product in human resulting from the catabolism of AMP is:*

A. Creatinine
B. Urea
C. Uric acid
D. Thiamin
E. Thymin

68. *A putrefactive waste product produced from tryptophan is:*

A. Serotonin
B. Melatonin
C. Cresol
D. Indican
E. Niacin

69. *A putrefactive waste product produced from tyrosine is:*

A. Serotonin
B. Melatonin
C. Cresol
D. Indican
E. Niacin

70. *The melanin pigments of hair and skin is derived from:*

A. Histidine
B. Tyrosine
C. Alanine
D. Arginine
E. Lysine

71. *Conjugation of glycine with benzoic acid produces:*

A. Hippuric acid
B. Glucuronic acid
C. Aspartic acid
D. GABA
E. Creatine

72. The main physiological methyl donor is:

A. Glutathione
B. S-adenosyl Methionine
C. Cysteine
D. Homocysteine
E. Serine

73. *Glutamate participates in synthesis of the following substances EXCEPT:*

A. Glutamine
B. Glutathione
C. Creatine phosphate
D. Folic acid
E. GABA

74. *Formic acid can be obtained from which of the following metabolic pathways:*
A. Urea cycle
B. Kreb's cycle
C. Tyrosine metabolism
D. Phenylalanine metabolism
E. Tryptophan metabolism

75. Fumaric acid can be obtained from the following EXCEPT:
A. Urea cycle
B. Kreb's cycle
C. Tyrosine metabolism
D. Phenylalanine metabolism
E. Tryptophan matabolism

In the following questions indicate with clear (T) the true statements, and with clear (F) the false statements:

Which of the following statements are TRUE for components of the urea cycle:
76. Urea is produced by the action of urease
77. The synthesis of carbamoylphosphate is stimulated by arginine amino acid
78. The reaction catalyzed by ornithine transcarbamoylase is the committed step of the cycle
79. Aspartate provides one of the urea nitrogens
80. The first 2 steps of the cycle occur in mitochondria

Pyridoxal phosphate is required for the enzymes catalyzing the following reactions:
81. Glutamate + NH_3 + ATP → Glutamine + ADP + Pi + H_2O
82. Glutamate + NAD^+ + H_2O → α-ketoglutarate + $NADH+H^+$ + NH_3
83. Glutamine + H_2O → Glutamate + NH_3
84. Glycine + Succinyl CoA → δ Amino levulonic acid
85. Glutamate + Pyruvate → α-ketoglutarate + Alanine

Tetrahydrofolate derivative is required for the conversion of:
86. Cobalamine to methyl cobalamine
87. Serine to glycine
88. Phenylalanine to tyrosine
89. Histidine to glutamate
90. Acetyl CoA to malonyl CoA

The conversion of propionyl CoA to succinyl CoA requires:
91. Tetrahydrofolate
92. Biotin
93. Vitamin B_{12}
94. More than one enzyme
95. GTP

S-Adenosyl Methionine (SAM) is a methylating agent in:
96. The conversion of norepinephrine to epinephrine
97. The conversion of phenylalanine to tyrosine
98. The synthesis of creatine from guanidoacetate
99. The synthesis of niacin from tryptophan
100. The synthesis dump to dTMP

In mammalian tissue, glycine is an important precursor on the pathway for the biosynthesis of:

101. Heme
102. Creatine
103. Catacholamine
104. Spermidine
105. Gaunine

For creatine:

106. It requires glycine, arginine and methionine for its synthesis
107. It acts as a carrier for long chain acyl CoA
108. Creatine phosphate contains high energy bond
109. It is synthesized in the kidney
110. Creatinine is anhydrous creatine

Which of the following statements about nitrogen metabolism are TRUE:

111. Formiminglutamate (FIGLU) is an intermediate in histidine degradation.
112. Cystathionine is cleaved to form serine and methione.
113. Glycine and arginine provide all the nitrogen for creatine synthesis.
114. S-Adenosylmethionine provides sulfur for biosynthesis of creatine phosphate.
115. Ammonia and Aspartate provide all nitrogen for urea synthesis.

The following α-amino acids have their corresponding α-ketoacids as intermediates of citric acid cycle:

116. Alanine
117. Aspartate
118. Valine
119. Glutamate
120. Ornithine

Chymotrypsin:

121. Is a gastric enzyme.
122. Acts on peptide bonds of small amino acids.
123. Its inactive form; chymotrypsinogen is activated by enteropeptidase enzyme.
124. Its secretion is stimulated by cholecystokinin.
125. It is exopeptidase.

The following α-amino acids are classified as essential amino acids for man:

126. Phenylalanine
127. Methionine
128. Glycine
129. Serine
130. Tryptophan

Pepsin

131. is secreted by body chief cells of the stomach.
132. is secreted as inactive form called trypsinogen.
133. its optimum pH for action is 1-2.
134. it is secreted under the influence of gastrin hormone.
135. is exopeptidase that acts on the periphery of polypeptide chains.

Glutathione:

136. Is synthesized in only one step catalyzed by glutathione synthase.
137. Has a role in amino acid absorption and transport.
138. Removes H_2O_2 in the presence of glutathione catalase enzyme.
139. is a tripeptide formed of arginine, cystine and aspartate.
140. inactivates insulin by hepatic insulin-glutathione transhydrogenase.

Aspartate:

141. Is glucogenic and ketogenic amino acid.
142. Acts as inhibitory transmitter in CNS.
143. Is a source of N1 of pyrimidine ring.
144. Gives asparagine by asparagine synthetase.
145. gives β-alanine by decarboxylation.

Creatine kinase enzyme:

146. has 3 isoenzymes MM, BB and MB.
147. catalyzes the conversion of creatine into creatinine.
148. Is present mainly in skeletal, cardiac muscles and brain.
149. Its level gets its peak after one week of the onset of myocardial infarction.
150. Is present in cardiac muscle mainly as MB isoenzyme.

Creatine:

151. is a simple protein.
152. Has a role of transport of long chain acyl CoA to the inside of the mitochondria for oxidation.
153. Is formed in kidney and liver and stored in skeletal muscles.
154. Its end product is creatinine.
155. Forms creatine phosphate that is the stored form of energy in muscle.

For the following amino acids:

A. *Aspartate*
B. *Methionine.*
C. *Glutamate.*
D. *Leucine.*
E. *Valine.*

156.
157. amino acids [A and B] are glucogenic.
158. amino acids [B, C and D] are essential.
159. amino acid [D is exclusively ketogenic.
160. amino acid [C] is degraded to pyruvate after transamination.
amino acid [E] is catabolized into succinyl CoA.

L-Glutamate dehydrogenase:

161.
162. its coenzyme is FAD
163. One of its functions is synthesis of non-essential amino acids.
164. It catalyzes the removal of amino group in the form of ammonia.
165. It is an enzyme present in most tissues.
It catalyzes an irreversible reaction.

Glycine

166.
167. is a precursor of thyroxin.
168. forms 1/3 of collagen protein.
169. ketogenic amino acid.
170. acts as inhibitory transmitter in spinal cord and medulla.
can be converted into serine by serine hydroxymethyl transferase.

In a 3-weeks old male infant with classical phenylketonuria:

171.
172. Tyrosine becomes an essential amino acid for the infant.
173. High levels of phenylalanine are found in blood.
174. The diagnosis of phenylketonuria is performed 4 days after birth.
175. A phenylalanine rich diet should be initiated immediately.
If untreated, the infant will develop mental retardation.

During the metabolism of branched chain amino acids:

176. Lipoic acid is not required
177. Leucine is converted to acetoacetyl CoA
178. Maple syrup urine disease results from deficiency of oxidative decarboxyltion of their α-ketoacids
179. Non of carbons of isoleucine is converted to succinyl CoA
180. Valine is pure glucogenic and is converted to succinyl CoA

Glutamate

181. Is provided in transamination reaction in which aspartate reacts with α-ketoglutarate.
182. Together with glutamine form half of amino acids pool of the body
183. Enters the urea cycle and directly provides carbon for synthesis of arginine
184. Is produced by the action of glutamate dehydrogenase on α-ketoglutarate
185. Acts as inhibitory neurotransmitter in brain.

Glutamate

186. Can provide both nitrogens of urea
187. Is often one of the products or reactants in transamination reactions
188. Can be oxdatively deaminated
189. Can be converted to glutamine in a single step
190. Can be converted to GABA in the brain

Serine:

191. Is the only amino acid that contains a hydroxyl group
192. Is a glucogenic amino acid
193. Is an essential amino acid
194. Is converted to glycine by a reaction requiring tetrahydrofolate
195. Is converted to pyruvate and ammonia by serine dehydratase

Matching: For each set of numbered questions, choose the ONE BEST answer from the list of lettered options below it. An answer may be used once or more times, or not at all.

196. Carnitine
197. Keratin
198. Creatine
199. Carnosine
200. Keratan sulphate

A. Glycosaminoglycans.
B. Methylgaunido acetic acid.
C. Conjugated histidine with β- alanine
D. Simple scleroprotein.
E. Denaturated protein
F. β-Hydroxy, γ trimethylamino butyric acid.

201. Glycine
202. Tryptophan
203. Serine
204. Aspartate
205. Arginine

A. Taurine
B. Sphingosine
C. β-Alanine
D. Nitric oxide
E. Heme
F. Niacin

206. Citrullinemia
207. Argininosuccinate aciduria
208. Argininemia
209. Hyperammonemia type I
210. Hyperammonemia type II

A. argininosuucinase
B. arginosuccinic acid synthase
C. Carbamoyl phosphate synthese I
D. arginase
E. Ornithine transcarbamoylase
F. Tryptophan pyrrolase

Match the numbered metabolic disorder with lettered deficient enzyme as they best fit together:

211. Albinism
212. Oxoprolinuria
213. Phenylketonuria
214. Homocystinuria
215. Hyperammonemia type II

A. Glutathione synthetase
B. Tyrosine hydroxylase
C. Cystathionine β- synthase
D. Carbamoyl phosphate synthase I
E. Phenylalanine hydroxylase
F. Ornithine transcarbamoylase
G. P-Hydroxyphenyl pyruvate oxidase

216. Endopeptidase acts on peptide bonds of basic amino acids
217. Endopeptidase acts on peptide bonds of aromatic amino acids
218. Endopeptidase acts on peptide bonds of small amino acids
219. Exopeptidase acts on peptide bonds of amino acids with free carboxylic group
220. Exopeptidase acts on peptide bonds of amino acids with free amino group

A. Rennin
B. Elastase
C. Trypsin
D. Gelatinase
E. Carboxypeptidase
F. Aminopeptidase
G. Pepsin

Match the numbered metabolic disorder with lettered deficient enzyme as they best fit together:

221. Hartnump's disease
222. Von Gierke's disease
223. Phenylketonuria
224. Albinism
225. Wilson disease

A. Glucose-6-phosphatase
B. Tryptophan pyrrolase
C. L-Glutamate dehydrogenase
D. Ceruloplasmin
E. Phenylalanine hydroxylase
F. Tyrosine hydroxylase

226. Alanine + ∝ -ketoglutarate → Pyruvate + Glutamate
227. Aspartate + α- ketoglutarate → Oxaloacetate + Glutamate
228. Arginine + O2 + NADPH+H^+ → Citrulline + NO + H2O + $NADP^+$
229. Ammonia +CO_2+2ATP → Carbamoyl phosphate
230. α-Ketoglutarate + NH3 → Glutamate
   A. Carbamoyl phosphate synthase I
   B. Glutamate dehydrogenase
   C. GGT
   D. AST
   E. ALT
   F. Nitric acid synthase

231. Creatine + ATP → Creatinine phosphate + ADP
232. Glycine + Methylene H4 folate ⇆Serine + H4 folate
233. Cysteine → Ethanolamine + CO_2
234. Phenylalanine + Tetrahydropterine+O2 → Tyrosine + dihydropterine+ H_2O
235. Tryptophan + Tetrahydrobiopterine → 5-hydroxytryptophan+ Dihydrobiopterine
   A. Tryptophan hydroxylase
   B. Phenylalanine hydroxylase
   C. Serine hydroxymethyl transferase
   D. Creatine kinase
   E. Cysteine decarboxylase

236. Produced as an inactive enzyme
237. Digest(s) dietary protein in stomach
238. Synthesized in the pancreas
239. Cleave(s) polypeptides at the carboxyl end of arginine and lysine residues
240. Optimal pH for activiy is 5
   A. Pepsin
   B. Trypsin
   C. Both of them
   D. Neither of them

241. Contain(s) carbon skeleton that can be converted to fumarate or acetoacetate
242. Is (are) essential in diet
243. Require(s) tetrahydrobiopterin for metabolism
244. Is (are) nonprotein amino acid(s)
245. Become(s) essential in absence of hydroxylase enzeme
   A. phenylalanine
   B. Tyrosine
   C. Both of them
   D. Neither of them

246. Required for the synthesis of epinephrine from norepinephrine
247. Required for the reaction catalyzed by phenylalanine hydroxylase
248. Required for the reaction catalyzed by ALT
249. Required for the transport of amino acids across cell membranes
250. Required for the reaction catalyzed by carbamoylphosphate synthase I
   A. N-Acetylglutamate
   B. Tetrahydrofolate
   C. Pyridoxal phosphate
   D. Biotin
   E. S-Adenosyl methionine
   F. CoA
   G. Tetrahydrobiopterin

251. Source of carbon 4, carbon 5 and nitrogen 7 of purines
252. May be converted to GABA
253. May be converted to dopamine
254. May be converted to glycine
255. May be converted to niacin

A. Tyrosine
B. Aspartate
C. Glutamate
D. Tryptophan
E. Glutamate
F. Serine
G. Glycine

256. Acetyl CoA and acetoacetate
257. Niacin
258. Carnitine
259. taurine
260. Spermidine

A. Arginine
B. Tryptophan
C. Histidine
D. Leucine
E. Ornithin
F. Cysteine
G. Lysine

261. Serotonin
262. γ-Aminobutyric acid
263. Hisitamin
264. Epinephrine
265. β-Alanine

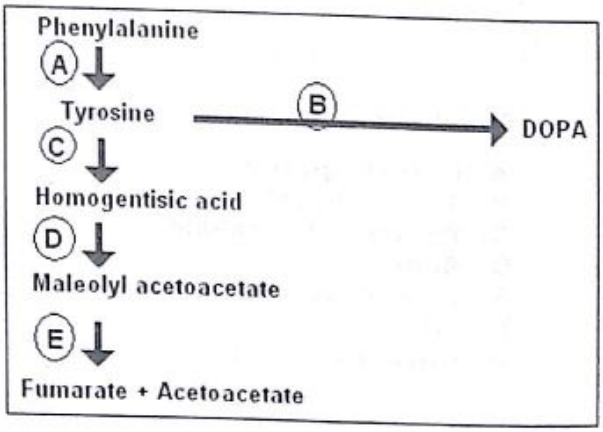
A. Glutamate
B. Cysteine
C. Tyrosine
D. Lysine
E. Histidine
F. Aspartate
G. Tryptophan

266. Taurine
267. Thyroxine
268. Ethanolamine
269. Acetoacetate
270. Glycoxylate

A. Tyrosine
B. Cysteine
C. Glycine
D. Alanine
E. Serine
F. Leucine
G. Ornithine

Match the numbered condition below with the lettered enzymatic defect in diagram at left:

271. Tyrosinosis
272. Albinism
273. Phenylketonuria
274. Alkaptonuria



Answer Key

MCQ:

1	2	3	4	5	6	7	8	9	10
B	A	D	D	A	B	C	C	B	D
11	12	13	14	15	16	17	18	19	20
B	A	D	A	B	E	D	C	C	B
21	22	23	24	25	26	27	28	29	30
A	B	C	B	B	A	B	E	C	D
31	32	33	34	35	36	37	38	39	40
A	A	A	D	D	C	D	E	E	C
41	42	43	44	45	46	47	48	49	50
A	C	C	B	C	C	B	E	A	A
51	52	53	54	55	56	57	58	59	60
C	A	B	C	C	C	A	E	D	A
61	62	63	64	65	66	67	68	69	70
D	A	D	B	B	C	C	D	C	B
71	72	73	74	75					
A	B	C	E	E					

True and false:

76	77	78	79	80	81	82	83	84	85
F	F	F	T	T	F	F	F	T	T
86	87	88	89	90	91	92	93	94	95
T	T	F	T	F	F	T	T	T	F
96	97	98	99	100	101	102	103	104	105
T	F	T	F	T	T	T	F	F	F
106	107	108	109	110	111	112	113	114	115
T	F	T	F	T	T	F	T	F	T
116	117	118	119	120	121	122	123	124	125
F	T	F	T	F	F	F	F	T	F
126	127	128	129	130	131	132	133	134	135
T	T	F	F	T	T	F	T	T	F
136	137	138	139	140	141	142	143	144	145
F	T	F	F	T	F	F	T	T	T
146	147	148	149	150	151	152	153	154	155
T	F	T	F	T	F	F	T	T	T
156	157	158	159	160	161	162	163	164	165
T	F	T	F	T	F	T	T	T	F
166	167	168	169	170	171	172	173	174	175
F	T	F	T	T	T	T	T	F	T
176	177	178	179	180	181	182	183	184	185
F	T	T	F	T	T	T	F	T	F
186	187	188	189	190	191	192	193	194	195
F	T	T	T	T	F	T	F	T	T

Matching:

196	197	198	199	200	201	202	203	204	205
F	D	B	C	A	E	F	B	C	D
206	207	208	209	210	211	212	213	214	215
B	A	E	C	F	B	A	E	C	F
216	217	218	219	220	221	222	223	224	225
C	G	B	E	F	B	A	E	F	D
226	227	228	229	230	231	232	233	234	235
E	D	F	A	B	D	C	E	B	A
236	237	238	239	240	241	242	243	244	245
C	A	B	B	D	A	A	A	D	B
246	247	248	249	250	251	252	253	254	255
E	G	C	C	A	G	C	A	F	D
256	257	258	259	260	261	262	263	264	265
D	B	G	F	E	G	A	E	C	F
266	267	268	269	270	271	272	273	274	
B	A	E	F	C	C	B	A	D	

Chapter 17

Biologic oxidation and Electron Transport Chain

1. *Thermodynamically, how reactions are classified?*

   A. Exothermic, isothermic and endothermic reactions.

2. *What is exothermic reaction?*

   A. Here energy is released from the reaction, and therefore reaction essentially goes to completion, e.g. urease enzyme, converting urea to ammonia + CO_2 + energy.

3. *What is endergonic reaction?*

   A. Energy is consumed and external energy is to be supplied for these reactions. In the body, this is usually accomplished by coupling the endergonic reaction with an exergonic reaction, e.g. Hexokinase reaction, Glucose + ATP $\Longrightarrow$ Glucose-6-Phosphate + ADP.

4. *What is the full name of ATP?*

   A. Adenosine triphosphate.

5. *What is the function of ATP?*

   A. It is the energy currency in the body. During the oxidation of foodstuffs, energy is released, a part of which is stored as chemical energy in the form of ATP. Other reaction requiring energy are coupled with ATP.

6. *Give examples of high energy compounds.*

   A. ATP, GTP, creatine phosphate, 1,3-bisphosphoglycerate, phosphoenol pyruvate, acetyl CoA, and succinyl CoA.

7. *On hydrolysis of 1 mole of ATP to ADP, the release of energy will be approximately how much?*

   A. 7 kCal.

8. *Where is respiratory chain located?*

   A. In the inner mitochondrial membrane.

9. *How is respiratory chain organised?*

   A. Components are organised into four complexes.

10. *What are the main activities taking place inside mitochondria?*

A. Citric acid cycle, electron transport chain, and β oxidation fatty acid.

11. *Where are enzymes of citric acid cycle located?*

A. Fluid matrix contains enzymes of citric acid cycle.

12. *What is the function of Co-enzyme Q?*

A. It catalyses the electron transport from complex I or II to complex III.

13. *Cytochrome oxidase is present in which complex?*

A. Complex IV.

14. *What are NAD+ linked dehydrogenases?*

A. Glyceraldehyde-3-phosphate dehydrogenase, pyruvate dehydrogenase, α-keto glutarate dehydrogenase, isocitrate dehydrogenase, malate dehydrogenase, β-hydroxy acyl CoA dehydrogenase.

15. *What are FAD-linked dehydrogenases?*

A. Succinate dehydrogenase, and acyl CoA dehydrogenase.

16. *Which cytochromes contain copper?*

A. Cytochrome oxidase.

17. *What is valinomycin?*

A. It acts as an ionophore, dissipates the proton gradient, and so inhibits ATP synthesis.

18. *What is malate-aspartate shuttle?*

A. It is a biochemical system for translocating electrons produced in cytosol during glycolysis into mitochondria.

19. *What are the salient features of ATP synthase?*

A. It has two subunits, Fo is a proton channel, and F1 has catalytic activity.

20. *How many ATP are produced in the oxidation of one molecule of NADH?*

A. Three.

21. *How many ATP are produced in the oxidation of one molecule of FADH?*

A. Two.

22. *Which is complex V of respiratory chain?*

A. ATP synthase.

23. *What is oligomycin?*

A. It inhibits oxidative phosphorylation.

24. *Name some inhibitors of oxidative phosphorylation.*

A. Oligomycin, carbon monoxide, and cyanide.

25. *Which inhibits electron transport chain at site I?*

A. Barbiturate.

26. *Which inhibits electron transport chain at site II?*

A. Antimycin A.

27. *Which inhibits electron transport chain at site III?*

A. Carbon monoxide and cyanide.

28. *What is the cause for death due to cyanide poisoning?*

A. Cyanide inhibits cytochrome oxidase.

29. *Which is a physiological uncoupler?*

A. Thyroxine.

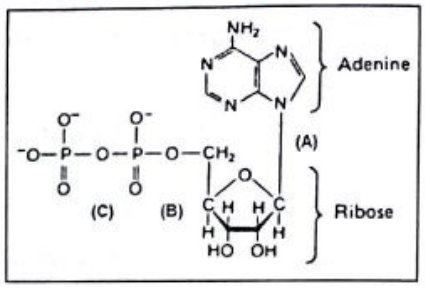
30. *How to explain the fever that accompanies calcium injection?*

A. Because calcium allows oxidation to proceed but prevents phosphorylation.

MCQ, Matching, True and False and Completion

Select and encircle the most appropriate answer or completion:

1. *In the diagram of ADP shown beside, three bonds are labeled A-B-C. Which is a high-energy bond?*



   A. A
   B. B
   C. C
   D. All of the above
   E. None of the above

2. *The free energy stored in the ATP molecule can be used for:*
   A. Chemical syntheses
   B. Heat, osmotic, and mechanical work
   C. Electrical work
   D. Driving endergonic reactions
   E. All of these

3. *The specific substrate for oxidative phosphorylation is:*
   A. AMP
   B. ADP
   C. ATP
   D. GTP
   E. $NADP^+$

4. *Which of the following activities occurs in mitochondria?*
   A. Cholesterol synthesis
   B. Acetoacetate synthesis
   C. Palmitate synthesis
   D. Splitting of arginine into urea and ornithine
   E. Insulin synthesis

5. *The effect of 2,4 dinitrophenol is to:*
   A. Raise the respiratory quotient
   B. Lower the respiratory quotient
   C. Decrease the H^+ gradient across the mitochondrial membrane
   D. Lower the basal metabolic rate
   E. Cause obesity

6. *In oxidative phosphorylation, the oxidation of one NADH+H+ to NAD+ produces how many ATPs?*
   A. 2
   B. 3
   C. 4
   D. 5
   E. 6

7. *Uncoupling of oxidative phosphorylation implies that:*
   A. The ATPase activity of mitochondria is abolished
   B. The mitochondria stop to oxidize succinate
   C. ATP formation stops but respiration continues
   D. ATP formation continues but respiration stops
   E. Cellular activities stops

8. Cytochromes are:
   A. Riboflavin-containing nucleotides
   B. Pyridine nucleotides
   C. Iron-porphyrin proteins
   D. Malate containing flavoproteins
   E. Peroxidation

9. *Entropy is a measure of the:*
   A. Rate of an enzymatic reaction
   B. Free energy of an enzymatic reaction
   C. Energy that is unavailable for work
   D. Exothermicity of a reaction
   E. Reversibility of a reaction

10. *Which of the following can serve as an inhibitor of electron transport?*
   A. Antimycin A
   B. Puromycin
   C. Actinomycin D
   D. Malonate
   E. Transhydrogenase

11. *If rotenone is added to the mitochondrial electron transport chain:*
   A. The P/O ratio of NADH + H^+ is reduced from 3:1 to 2:1
   B. The rate of NADH + H^+ oxidation is diminished to 2/3 of the initial value
   C. Succinate oxidation remains normal
   D. Oxidative phosphorylation is uncoupled at site I
   E. Electron flow is inhibited at site II

12. *Cytochrome oxidase contains:*
   A. Cobalt
   B. Zinc
   C. Magnesium
   D. Selenium
   E. Copper

13. *If cyanide is added to tightly coupled mitochondria that are actively oxidizing succinate:*
   A. Subsequent addition of 2,4 dinitrophenol will cause ATP hydrolysis
   B. Subsequent addition of 2,4 dinitrophenol will restore succinate oxidation
   C. Electron flow will cease , but ATP synthesis will continue
   D. Electron flow will cease , but ATP synthesis can be restored by subsequent addition of 2,4 dinitrophenol
   E. Subsequent addition of 2,4 dinitrophenol and the phosphorylation inhibitor oligomycin will cause ATP hydrolysis

14. *In substrate level phosphorylation*
   A. The substrate reacts to form a product containing a high energy bond
   B. ATP synthesis is linked to dissipation of a proton gradient
   C. High energy intermediate compounds cannot be isolated
   D. Oxidation of one molecule of substrate is linked to synthesis of more than one ATP molecule
   E. Mitochondria participate, but not cytosol

15. *Which cytochrome is involved in direct reduction of oxygen?*
   A. Cytochrome a_1
   B. Cytochrome b_5
   C. Cytochrome C_1
   D. Cytochrome d
   E. Cytochrome oxidase

In the following questions indicate with clear (T) the true statements, and with clear (F) the false statements:

Oxidative phosphorylation:

16. Is the formation of ATP in association with oxidative processes.
17. Is the formation of ATP by ATP synthase present in the inner mitochondrial membrane.
18. Is said to be "uncoupled" when substrate and not ADP is phosphorylated.
19. Is said to be "uncoupled" when electron transport proceeds without formation of ATP.
20. Is stimulated by calcium ions.

The chemiosmotic hypothesis involves:

21. A membrane impermeable to proton.
22. Electron transport by the respiratory chainpumps protons out of the mitochondria
23. Proton flow into the mitochondria depends on the presence of ADP and Pi.
24. ATPase activity is reversible
25. Only proton transport is strictly regulated; other positively charged ions can diffuse freely across the mitochondrial membrane.

Matching: For each set of numbered questions, choose the ONE BEST answer from the list of lettered options below it. An answer may be used once or more times, or not at all.

26. *Catalyzes electron transport between flavoprotein and cytochrome b*
27. *terminates the electron transport chain*
28. *Catalyzes electron transport between ubiquinone and cytochrome C*
29. *A constituent of cytochrome oxidase*
30. *Catalyzes electron transport between NADH+H$^+$ and ubiquinone*
   A. Ubiquinone
   B. Flavoprotein
   C. Cytochrome oxidase
   D. Cytochrome a
   E. Cytochrome b

31. *Electron transport*
32. *Hemoprotein*
33. *Flavoprotein*
34. *Cytochrome oxidase*
35. *High energy compound*

A. Ubiquinone
B. Cytochrome
C. Both Ubiquinone and cytochrome c
D. Neither ubiquinone nor cytochrome c

Answer Key

MCQ:

1	2	3	4	5	6	7	8	9	10
C	E	B	B	C	B	C	C	C	A
11	12	13	14	15					
C	E	A	A	E					

True and false:

16	17	18	19	20	21	22	23	24	25
T	T	F	T	F	T	T	T	T	F

Matching:

26	27	28	29	30	31	32	33	34	35
A	C	E	D	B	C	B	D	D	D

Chapter 18

Heme Synthesis and Breakdown

1. *What are heme containing compounds (hemoproteins)?*

   A. Hemoglobin, myoglobin, respiratory cytochromes, cytochrome p450, catalase, peroxidase, and tryptophan oxygenase.

2. *What are iron containing proteins?*

   A. Hemoglobin, myoglobin, respiratory cytochromes and cytochrome p450.

3. *What are the functions of hemoglobin and myoglobin?*

   A. Hemoglobin carries oxygen from lungs to tissues and myoglobin stores oxygen in muscles.

4. *What are the structural features of hemoglobin A molecule?*

   A. Hb A has four subunits, two α and two β units. It contains 4 iron atoms.

5. *How many molecules of oxygen can bind with hemoglobin?*

   A. Four.

6. *What are the structural features of myoglobin molecule?*

   A. It has one subunit, and contains one iron atom.

7. *How many molecules of oxygen can bind with myoglobin?*

   A. One.

8. *100 ml of blood can carry how much oxygen?*

   A. 20 ml.

9. *What is Bohr effect?*

   A. It is the influence of pH and pCO2 to facilitate oxygenation of Hb in the lungs and deoxynation at the tissues.

10. *What is cooperative binding of O2?*

    A. Binding of one oxygen molecule at one heme, increases the oxygen affinity of the remaining heme groups in the same hemoglobin molecule.

11. *Which will decrease the affininty of hemoglobin for oxygen?*

    A. 2,3 Bisphosphoglycerate (2,3-BPG) level.

12. What is chloride shift?

A. Bicarbonate ions diffuse outside the RBCs in exchange for chloride ions which shifts into the cell in order to maintain electrical neutrality across the erythrocyte membrane.

13. What are diferent types of hemoglobin?

A. Hemoglobin A, hemoglobin A_2, fetal hemoglobin (HbF), and hemoglobin A_{1c} (glycated hemoglobin).

14. What is the structural difference between HbA, HbA_2 and HbF?

A. HbA has 2 α and 2 β chains, HbA_2 has 2 α and 2 δ chains and HbF has 2 α and 2 γ chains.

15. What is hemoglobin A_{1c} (glycated hemoglobin)?

A. It is hemoglobin A + Glucose. Glucose is added non enzymatically to hemoglobin A. The higher the blood glucose, the higher the the precent of glycated hemoglobin.

16. What is the importance of measuring hemoglobin A_{1c}?

A. High percent of hemoglobin A1c indicate bad diabetic control over the past 2 months.

17. What are the laboratory tests to identify HbF?

A. By electrophoresis. HbF moves slower than HbA on electrophoresis.

18. What is the physiological significance of HbF?

A. Oxygen affinity is more for HbF than HbA. HbF is seen in fetal circulation.

19. May Hb F be seen in adult blood?

A. Yes it may be seen in adults in cases of hemoglobinopathies and thalassemias.

20. Why carbon monoxide becomes a poison?

A. Hb has 200 times more affinity to carbon monoxide than oxygen.

21. What is the treatment for carbon monoxide poison?

A. Hyperbaric oxygen.

22. Met-hemoglobin is found in which conditions?

A. Ingestion of nitrites, presence of HbM, glucose -6- phosphate dehydrogenase deficiency.

23. What is met-hemoglobin?

A. Hemoglobin in which iron is in ferric state.

24. What is the defect of met-hemoglobin?

A. It cannot release oxygen in tissues.

25. What is the reagent used for colourimetric estimation of hemoglobin?

A. Drabkin's reagent, to convert Hb to cyanmet-hemoglobin.

26. *What is hemoglobin S?*

A. The glutamic acid in the 6th position of β chain of HbA is changed to valine in HbS.

27. *What is the cause for sickle cell anemia?*

A. Solubility of deoxy HbS is lower than deoxy HbA, so HbS is precipitated intracellularly, leading to sickle shape of RBC.

28. *How HbS is identified?*

A. By its characterstic shape in blood films and HbS is slower moving on electrophoresis than HbA.

29. *What is sickle cell trait?*

A. Heterozygous condition, one allele is normal, the other is abnormal, so half of Hb molecules are normal, and half abnormal.

30. *What is its clinical significance?*

A. Usually sickle cell trait persons will not have any disease manifestations, but, at higher altitudes, hypoxia may cause manifestation of the disease. Chronic lung disorders may also produce hypoxia-induced sickling in HbS trait.

31. *What is hemoglobin C?*

A. The glutamic acid in the 6th position of β chain of HbA is changed to lysine in HbC. It is less harmfull compared to HbS.

32. *What is the chemical structure of heme?*

A. It is ferro protoporphyrin.

33. *What are the substituent groups of heme?*

A. Methyl, vinyl, and propionyl groups.

34. *Heme is synthesised from what substances?*

A. It is synthesised from glycine and suucinyl CoA.

35. *What is the rate limiting enzyme (key enzyme) of heme synthesis?*

A. ALA synthase.

36. *ALA synthase present in which cell organelle?*

A. Mitochondria.

37. *What is the coenzyme for ALA synthase?*

A. Pyridoxal phosphate.

38. *How is heme synthesis regulated?*

A. ALA synthase is feed back inhibited by heme, and stimulated by certain drugs as phenobarbital and iron.

39. *What is the action of barbiturates on heme synthesis?*

A. Barbiturates stimulate heme synthesis.

40. *Which enzyme is inhibited by lead?*
A. ALA dehydratase and ferrochelatase.

41. *Methenyl bridge of protoporphyrin is derived from what?*
A. α carbon of glycine.

42. *What are porphyrias?*
A. These are a group of diseases resulting from a deficiency of one of the enzymes needed for heme synthesis.

43. *What are types of porphyrias?*
A. Acute intermittent porphyria, porphyria cutanea tarda, hereditary coproporphyria, variegate porphyria, congenital erythropoietic porphyria, and protoporphyria.

44. *What is the enzyme defect in acute intermittent porphyria?*
A. Uroporphyrinogen I synthase.

45. *In acute intermittent porphyria, urine contains what?*
A. ALA and PBG.

46. *What is the enzyme defect in porphyria cutanea tarda?*
A. Uroporphyrinogen decarboxylase.

47. *What is the enzyme defect in hereditary coproporphyria?*
A. Coproporphyrinogen oxidase.

48. *What is the enzyme defect in variegate porphyria?*
A. Protoporphyrinogen oxidase.

49. *What is the enzyme defect in congenital erythropoietic porphyria?*
A. Uroporphyrinogen III synthase.

50. *What is the enzyme defect in protoporphyria?*
A. Ferrochelatase

51. *What are effects of porphyrias?*
A. Anemia, abdominal pain, neuropsychatric symptoms and photosensitivity.

52. *What is the cause of photosensitivity?*
A. Some porphyrin derivatives when exposed to light react with molecular oxygen to form oxygen radicals, which cause skin damage.

53. *Lead poisoning results in what?*
A. acquired porphyria.

54. *Lead poisoning results in elevated levels of what?*
A. Delta amino levulinic acid.

55. *Degradation of heme needs which enzyme?*
A. Heme oxygenase system, with NADPH.

56.Is carbon monoxide produced by the body?

A. Yes. Through degradation of heme to bilirubin.

57.Where does extravascular hemolysis occur?

A. In reticulo-endothelial cells mostly in liver, spleen and bone, for RBCs at the end of their life span (120 days).

58.What is the end product of heme catabolism?

A. Bilirubin.

59.What is the function of bilirubin?

A. It may act as antioxidant.

60.Heme is converted to bilirubin in which site?

A. Microsomes of reticulo-endothelial cells.

61.Bilirubin in blood is carried by what?

A. Albumin.

62.How is it made water soluble?

A. By conjugation with glucoronic acid.

63.Where is the conjugation taking place?

A. In liver.

64.What is the enzyme?

A. UDP-glucuronyl transferase.

65.What is the normal level of total plasma bilirubin?

A. 0.2 - 1.0 mg/dl.

66.What is the normal level of conjugated bilirubin in plasma?

A. Less than 0.2 mg/dl.

67.What is jaundice?

A. When plasma bilirubin is more than 3 mg/dl, it diffuses into tissues,causing yellowish discolouration of tissues.

68.What is latent jaundice?

A. When plasma bilirubin is between 1 to 3 mg/dl.

69.What is the difference between hyperbilirubinemia and jaundice?

A. Hyperbilirubinemia is a laboratory term means increased plasma bilirubin above normal level (1.2 mg/dl), where jaundice, is a clinical term, characterized by yellow discoloration of skin, sclera and mucus membrane. It is due to increase plasma bilirubin above 3 mg/dl.

70.Enterohepatic circulation is seen in which substances?

A. Urobilinogen and bile salts.

71.Bilirubin in serum is estimated by what test?

A. Van den Bergh reaction.

72. *What is direct van den Berg's reaction?*

A. The colour is developed immediately when blood is added.

73. *What is your interpretation,when direct test is positive?*

A. Blood contains conjugated bilirubin,it is water soluble.

74. *What is indirect Van Den Bergh's test?*

A. When blood is added to the solution, there is no colour, but when alcohol is added, colour is developed.

75. *What is the reason for this type of reaction?*

A. Bilirubin is soluble in alcohol, and alcohol extract gives the reaction.

76. *In obstructive jaundice, what is seen in blood?*

A. Conjugated bilirubin in excess quantity.

77. *Bile salts and bile pigments are excreted in urine in which condition?*

A. Obstructive jaundice.

78. *What is the reason for characterstic brown color of stool?*

A. Urobilinogen (=strecobilinogen).

79. *In obstructive jaundice, what is the color of stool?*

A. Clay color due to absence of urobilinogen.

80. *What are causes of most cases of obstructive jaundice?*

A. Gall stones and cancer head of pancreas.

81. *In hepatocellular jaundice, what is seen in blood?*

A. Excess conjugated and unconjugated bilirubin.

82. *What are causes of hepatocellular jaundice?*

A. It is due to liver cells damage as in cirrhosis, viral hepatitis or toxins.

83. *Increased urobilinogen in urine is seen in which conditions?*

A. In all cases of hemolytic anemias as congenital spherocytosis, mismatched transfusion, Rh incompatability, and glucose-6-phosphate dehydrogenase deficiency.

84. *What is neonatal jaundice?*

A. This is a transient condition occurs in some newborn infants especially if they are premature.

85. *What are causes of neonatal jaundice?*

A. At birth, liver contains very little UDP-glucuronyltransferase enzyme, which is important for conjugation of bilirubin. This leads to accerelated hemolysis of RBCs.

86. *What is kernictrus?*

A. In young children, when plasma bilirubin is more than 20 mg/dl, it diffuses into brain, causing permanent damage to brain cells.

87.What are causes of congenital hyperbilirubinemia?

A. Gilbert's disease, Cligler-Najjar syndrome and Dubin - johnson syndrome.

88.What is the defect in Gilbert's disease?

A. Uptake of bilirubin by the liver is defective.

89.What is the defect in Ciggler-Najjar syndrome?

A. Defect in conjugation of bilirubin due to deficiency of UDP glucuronyl transferase.

90.What is the defect in Dubin - johnson syndrome?

A. hepatic secretion of conjugated bilirubin into the bile.

91.Compare between unconjugated and conjugaterd bilirubin.

	Unconjugated (indirect) bilirubin	Conjugated (direct) bilirubin
Site	Present normally in plasma	Present normally in bile
Conjugation	Unconjugated, attached noncovallently to albumin	Conjugated to glucuronic acid.
Molecular weight	Has higher molecular weight, and cannot be filtered through kidney.	Has smaller molecular wight, and can be filtered through kidney.
Solubility	Nonpolar, insoluble in plasma and can cross brain barrier in neonates causing brain damage.	Polar, soluble in plasma and cannot cross brain barrier.
Van Den Bergh reaction	Indirect	Direct

92.Compare between different types of porphyrias.

Types and class	Enzyme involved	Major symptoms
Hepatic porphyrias:		
Acute intermittent porphyria	Uroporphyrinogen I synthase	Abdominal pain Neuropsychatric
Porphyria cutanea tarda	Uroporphyrinogen decarboxylase	Photosensitivity
Hereditary coproporphyria	Coproporphyrin oxidase	Abdominal pain Neuropsychatric Photosensitivity
Variegate porphyria	Protoporphyrinogen oxidase	Abdominal pain Neuropsychatric Photosensitivity
Erythropoietic porphyrias:		
Congenital erythropoietic porphyrias:	Uroporphyrinogen III synthase	Photosensitivity
Erythro hepatic porphyrias:		
Protoporphyria	Ferrochelatase	Photosensitivity

MCQ, Matching, true and false and Completion

Select and encircle the most appropriate answer or completion:

1. *All of the followings are normal hemoglobin EXCEPT:*
   A. Hemoglobin A_1
   B. Hemoglobin A_{1c}
   C. Hemoglobin S
   D. Hemoglobin A_2
   E. Hemoglobin F

2. *Carbon monoxide produce hypoxia by:*
   A. Forming met-hemoglobin
   B. Occupying the same position of O_2 in the hemoglobin
   C. Removing iron form hemoglobin, forming hematin
   D. Slowing capillary circulation
   E. Inhibiting mitochondrial oxidative phosphorylation

3. *All the following proteins contain porphyrin ring EXCEPT:*
   A. Hemoglobin
   B. Myoglobin
   C. Cytochrome
   D. Catalase
   E. Peroxidase

4. *The following disorders are inherited as autosomal dominant diseaseEXCEPT:*
   A. acute intermittent porphyria.
   B. hereditary coproporphyria.
   C. porphyria variegate.
   D. Congenital erythropoietic porphyria.
   E. Erythrohepatic porphyria

5. *The first step in the catabolism of hemoglobin occurs when hemoglobin is:*
   A. Converted to biliverdin in the liver
   B. Converted to bilirubin in the reticuloendothelial cells
   C. Conjugated with glucuronic acid in the liver
   D. Reduced in the liver
   E. Reduced in the intestine

6. *Delta bilirubin is that type of bilirubin which:*
   A. Present normally in blood.
   B. Present normally in bile.
   C. Present in trace amounts in urine
   D. Binds noncovalently with albumin.
   E. Binds covalently with albumin

7. *Thalassemia is an example of:*

A. Frame shift mutation.
B. Seilent mutation.
C. Missense mutaion.
D. Nonsense mutation.

8. *Bile pigments are derived from:*

A. Cholic acid
B. Heme
C. Triacylglycerols
D. Cholesterol.
E. Phospholipids

9. *All about δ - Aminolevulonic acid synthase activity is true EXCEPT:*

A. Catalyzing the rate limiting reaction in heme biosynthesis.
B. Requires the coenzyme pyridoxal phosphate.
C. Occurs in mitochondria.
D. Its substrates are acetyl CoA and glycine.

10. *Bilirubin diglucuronide is:*

A. Elevated in neonatal hyperbilirubinemia.
B. Elevated in hemolytic jaundice.
C. Lipid soluble
D. Normally excreted in urine
E. Usually found in bile duct

11. *In heme synthesis, the committed step is:*

A. Condensation of 2 PBG (porphobilinogen).
B. Condensation of 2 ALA (δamino levulonate).
C. Condensation of glycine and succinyl CoA.
D. Formation of uroporphyrinogen III.
E. Formation of coproporphyrinogen III

12. *The type of hemoglobin which binds irreversibly with oxygen is:*

A. Hemoglobin A
B. Hemoglobin A2.
C. Hemoglobin F.
D. Hemoglobin S.
E. Methemoglobin

13. *All the followings are types of congenital hyperbilirubinemia EXCEPT:*

A. Gilbert's disease
B. Von Gierke's disease
C. Crigler Najjar Syndrome
D. Dubin Johnson Syndrome

14. *δ - Aminolevulonic acid synthase activity:*

A. Its activity is repressed in individuals treated with drugs such as phenobarbital.
B. Catalyzing the rate limiting reaction in heme biosynthesis.
C. Requires the coenzyme biotin.
D. Occurs in cytosol.
E. Is strongly stimulated by heavy metals as lead.

15. *The catabolism of hemoglobin involves:*
A. The formation of bile pigments.
B. Oxidative cleavage of porphyrin ring.
C. The formation of urobilin.
D. Results in liberation of carbon monooxide.
E. All of the above.

16. *Erythrocytes of peoples with a homozygous sickle cell anemia contain predominantly:*
A. Hemoglobin A.
B. Hemoglobin A2.
C. Hemoglobin F.
D. Hemoglobin S.
E. Methemoglobin

17. *The normal brown –red color of feces results from the presence of:*
A. Heme.
B. Stercobilin.
C. Bilirubin diglucuronide.
D. Biliverdin.
E. Coproporphyrin III.

18. *All of the following statements are correct EXCEPT:*
A. In normal individuals bilirubin is not detectable in urine.
B. Hemolysis elevates plasma unconjugated bilirubin.
C. "Direct –reacting" bilirubin in the Van den Bergh reaction corresponds to conjugated bilirubin.
D. Hemolytic jaundice results in decreased excretion of urinary urobilinogen.

19. *In sickle cell hemoglobin the position 6 glutamyl residue of the β-chain of normal adult hemoglobin is replaced by:*
A. Aspartate.
B. Threonine.
C. Lysine.
D. Glutamine.
E. Valine

20. *The direct bilirubin consists of:*
A. Bilirubin conjugated with glucuronic acid
B. Bilirubin conjugated with gluconic acid
C. Bilirubin conjugated with albumin
D. Bilirubin conjugated with globulin

21. *In hemoglobin C the position 6 glutamyl residue of the β-chain of normal adult hemoglobin is replaced by:*
A. Aspartate.
B. Threonine.
C. Lysine.
D. Glutamine.
E. Valine

22. *Metabolic degradation of hemoglobin takes place mainly in:*
A. The reticulendithelial system.
B. The erythrocytes.
C. The skeletal muscles.
D. Kidney tubules.
E. All the above.

23. *Which is not part of hemoglobin molecule?*
   A. Histidine.
   B. Vinyl group.
   C. Frric ion
   D. Pyrrole rings.
   E. Protein.

24. *Which of the following amino acid is the precursor of δ - amino levulonate:*
   A. Alanine.
   B. Proline.
   C. Glycine.
   D. Leucine.
   E. Histidine.

25. *The Bohr effect describes:*
   A. The effect of PCO_2 on the dissociation of oxyhemoglobin.
   B. The effect of zn^{2+} on the activity of carbonic anhydrase.
   C. The toxic effect of very high PO_2 on the nervous system.
   D. The buffering action of imidazole groups.
   E. All of the above.

26. *Crigler Najjar syndrome is a congenital bilirubinemia due to a defect in the conjugating system due to the lack of which enzyme:*
   A. UDP-glucose epimerase.
   B. Phosphogalactose-uridyl transferase.
   C. UDP-glucuronyl transferase.
   D. Phosphoglucose-uridyl transferase.
   E. UDP-galactose pyrophosphoryl

In the following questions indicate with clear (T) the true statements, and with clear (F) the false statements:

In the synthesis of heme:

27. *The First reaction is the condensation of glycine and succinate.*
28. *Porphobilinogen (PBG) is formed from two molecules of ALA.*
29. *Coproporphyrinogen is synthesized before uroporphyrinogen.*
30. *Iron is incorporated into protoporphyrin.*
31. *The rate limiting step is catalyzed by ALA-synthase.*

Recognized major symptoms of the inherited porphyries include:

32. *Abdominal pain.*
33. *Neuropsychiatric disorders.*
34. *Liver cell failure.*
35. *Photosensitivity of the skin*
36. *Diarrhea.*

In porphyries:

37. *In the porphyria cutanea tarda. the deficient enzyme is uroporphyrinogen I synthase.*
38. *In porphyria variegate abdominal pain, neuropsychiatric disorders and photosensitivity are present.*
39. *In hereditary coproporphyria, the deficient enzyme is coproporphyrin oxidase.*
40. *In acute intermittent porphyria neuropsychiatric disorders do not occur.*
41. *Congenital erythropoietic porphyria is autosomal dominant.*

ALA synthase activity is increased:

42. By increased level of heme.
43. By the administration of barbital.
44. By the administration of glucose.
45. By the administration of steroids.
46. By the administration of iron.

Matching: For each set of numbered questions, choose the ONE BEST answer from the list of lettered options below it. An answer may be used once or more times, or not at all.

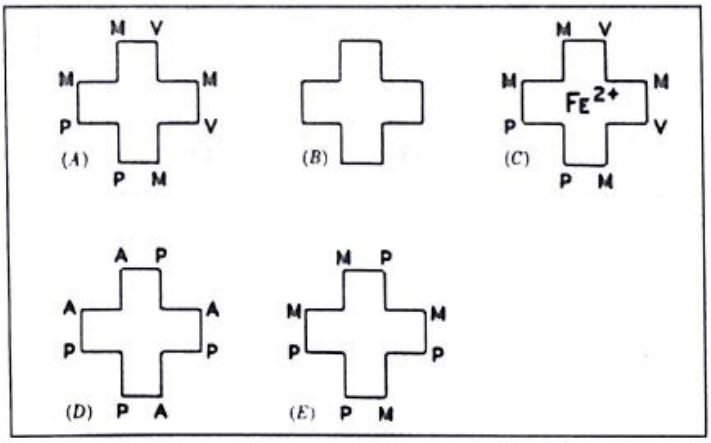
47. *Hepatic cirrhosis*
48. *Acute attack of sickle cell anemia*
49. *Neonatal Jaundice*
50. *Crigler Najjar syndrome*
51. *Obstructive jaundice*

A. Serum direct bilirubin is elevated
B. Serum indirect bilirubin is elevated
C. Both are elevated
D. Neither are elevated

52. *Acute Intermittent porphyria*
53. *Porphyria cutanea tarda*
54. *Hereditary coproporphyria.*
55. *Variegate porphyria*
56. *Protoporphyria.*

A. Coproporphyrin oxidase.
B. Uroporphyrinogen decarboxylase.
C. Uroporphyrinogen I synthase.
D. Ferrochelatase.
E. Protoporphyrinogen oxidase.

57. *Coproporphyrin III.*
58. *Heme.*
59. *Protoporphyrin type III.*
60. *Porphin.*
61. *Uroporphyrin III.*



Answer Key

MCQ:

1	2	3	4	5	6	7	8	9	10
C	B	E	D	A	E	A	B	D	E
11	12	13	14	15	16	17	18	19	20
B	E	B	B	E	D	B	D	E	A
21	22	23	24	25	26				
C	A	C	C	A	C				

True and false:

27	28	29	30	31	32	33	34	35	36
T	T	F	T	T	T	T	F	T	F
37	38	39	40	41	42	43	44	45	46
F	T	T	F	F	F	T	F	F	T

Matching:

47	48	49	50	51	52	53	54	55	56
C	B	B	B	A	C	B	A	E	D
57	58	59	60	61					
E	C	A	B	D					

Chapter 19 *Immunochemistry*

1. *What are immunoglobulins?*

   A. These are a group of proteins (gamma globulins) produced by the β-lymphocytes and plasma cells in response to the presence of foreign compounds (antigens).

2. *How immunoglobulins are classified?*

   A. IgM , IgA , IgG , IgE, and IgD.

3. *Which is more common in plasma?*

   A. IgG (80%).

4. *Which one can cross the placental barrier?*

   A. IgG.

5. *What is the basic structure of immunoglobulins?*

   A. Two heavy chains and two light chains connected by disulphide linkages.

6. *Describe the structure of immunoglobulin.*

   A. Both heavy and light chains are glycoproteins. Each has 2 regions, C-terminal constant region, which has a constant amino acid sequences within a class or type and N-terminal variable (v) regions, with considerable variation in amino acid sequence from molecule to another.

7. The antigen binding capacity of immunoglobulin resides at which region of immunoglobulin?

   A. Variable region.

8. *How many binding sites per immunoglobulin molecule?*

   A. 2 per immunoglobulin molecule, a property known as divalency.

9. *What are types of light chains?*

   A. There are only 2 major types in man, the Kappa, κ (70%) and Lambda, λ (30%). Either types of light chain can be associated with each of the heavy chain classes.

10. What is primary immune response antibody?

    A. Immunoglobulin M.

11. *What do you mean by primary immune response?*

A. It is the first of the antibodies, which act on introduction of a foreign antigen into the plasma. Its presence indicates recent infection.

12. *Which immunoglobulin having the highest molecular weight?*

A. IgM as it is formed of 5 basic subunits (pentamer).

13. *What is the secondary* immune *response antibody?*

A. Immunoglobulin G.

14. *Which antibody is seen in body secretions?*

A. Immunoglobulin A.

15. *Which immunoglobulin has a secretory piece?*

A. Immunoglobulin A.

16. *What is the function of secretory piece?*

A. It protects IgA from digestion by gastrointestinal proteolytic enzymes.

17. *What is the function of IgA?*

A. It protects body surfaces against invading microorganisms, (in the intestinal, respiratory, and urogenital tracts as well as milk, colostrums and tears).

18. *What is the clinical importance of immunoglobulin E?*

A. They mediate allergic (anaphylactic) reactions.

19. *What is the cause of anaphylactic reaction?*

A. IgE (through its Fc regions), in the presence of antigen, binds to mast cells, leading to mast cell degranulation and release of histamine and other substances. These substances result in allergic manifestations

20. *Which immunoglobulin having shortest half life?*

A. Immunoglobulin E.

21. *What is M band?*

A. A narrow peak in gamma globulin, caused by monoclonal antibodies secreted by malignant plasma cells.

22. *What is Bence Jones protein?*

A. It is the light chains of immunoglobulins, excreted in urine. It is seen in urine of 20% cases of multiple myeloma.

23. *What is the test done to detect Bence jones protein?*

A. Bence jones protein in urine is precipitated when heated at 60°C, dissolved at 100°C and re-precipitated again when it is cooled to 60°C.

24. *How antibody diversity is produced?*

A. By somatic recombination.

25. *What is antigen?*

A. Are substances when introduced into the body will stimulate an immune response i.e. antibodies production.

26. What are *haptens?*

A. Are small molecules that can not by themselves induce antibody formation but can do so when covalently linked to larger molecules.

27. What is *antigen determinant?*

A. This is the portion of antigen that binds with antigen receptors.

28. What are *antigen receptors?*

A. These are the sites on the surface of lymphocytes that bind the antigen.

29. What is *immunopotency?*

A. It is the capacity of the region of the antigenic determinant to induce the formation of specific antibodies.

30. *What are properties of antigens?*

A. They have high molecular weight and structural complexity. They are foreign to the body. The protein with molecular weights greater than 100,000 are the most potent antigens.

31. *May body tissues and body proteins stimulate immune response?*

A. No, because under normal conditions, tissues or fluids of the body can be recognized by immune system as self (i.e. own tissues), and do not stimulate immune response.

32. *What happen if tissues of the body stimulate immune system?*

A. This leads to autoimmune diseses as rheumatoid arthiritis.

MCQ, Matching, true and false and Completion

Select and encircle the most appropriate answer or completion:

1. *Which statement about Immunoglobulin G (IgG) is CORRECT:*
   A. The only immunoglobulin that can pass the placenta.
   B. Forms 20 % of the total immunoglobulins.
   C. Present exclusively in intestinal secretions.
   D. Immunoglobulins of primary immune response
   E. Contains secretory components.

2. *An antigen is characterized by all the followings EXCEPT:*
   A. Having regions of sequence hypervariability
   B. Protein in nature
   C. Having a high molecular weight
   D. Foreign to the host
   E. Having a complex structure

3. *The immunoglobulin that passes through the placenta is:*
   A. IgA
   B. IgD
   C. IgE
   D. IgG
   E. IgM

4. *Which statement about Immunoglobulins is INCORRECT:*
   A. Are associated with γ globulins fraction by electrophoresis.
   B. Their basic structure has 4 polypeptide chains.
   C. Are synthesized mainly in the liver.
   D. Not all of them can pass the placenta.

5. *Immunoglobulin M (IgM):*
   A. Has a relative long half life.
   B. Is Present as monomer or dimer.
   C. The major immunoglobulin during primary immune response.
   D. Can cross the placenta.

6. *Haptens:*
   A. Can alone stimulate antibody production.
   B. Large molecules having structural complexity.
   C. They have no antigenic determinant.
   D. It is antigenic after covalently bound to large molecules.

7. *IgG:*
   A. Is found primarily in mucosal secretion.
   B. Is one of less common immunoglobulins.
   C. Contains carbohydrate covalently attached to the H chain.
   D. It helps the release of histamine by mast cells.

8. *In immunoglobulins all of the following are true EXCEPT:*
   A. There are four polypeptide chains.
   B. There are 2 copies of each type of heavy and light chains.
   C. All chains are linked by disulfide bonds.
   D. Carbohydrate is covalently bound to the protein.
   E. Immunoglobulin class is determined by the light chains.

9. *Which statement about antigens is INCORRECT:*
   A. Is foreign to the body
   B. They cannot by themselves induce antibodies formation.
   C. Have high molecular weight.
   D. Posses some degree of complexity.

10. *Bence Jones protein*
   A. It is the heavy chains of immunoglobulins.
   B. It is precipitated at 20°C.
   C. It is seen in urine of 20% cases of multiple myeloma.
   D. It is protein normally excreted in urine.

Matching: For each set of numbered questions, choose the ONE BEST answer from the list of lettered options below it. An answer may be used once or more times, or not at all.

11. *Immunoglobulin A*
12. *Immunoglobulin M*
13. *Immunoglobulin D*
14. *Immunoglobulin E*
15. *Immunoglobulin G*
   A. It has activity against thyroid tissue, insulin and penicillin.
   B. Constitutes 80% of all serum immunoglobulins.
   C. It has polysaccharide nature.
   D. Major immunoglobulin during the primary immune response.
   E. contains secretory piece.
   F. Formed of kappa chains only.
   G. Responsible for immunity against parasites.

Answer Key

MCQ and matching:

1	2	3	4	5	6	7	8	9	10
A	A	D	C	C	D	C	E	B	C
11	12	13	14	15					
E	D	A	G	B					

Chapter 20 *Body fluids*

1. *What is the composition of blood?*

   A. Blood is a liquid consisted of a yellowish fluid called plasma in which red and white blood cells and platelets are suspended.

2. *What is normal serum albumin level?*

   A. 3.5 - 5 g/dl.

3. *What is dl?*

   A. Deciliter = 100 ml.

4. *What is the normal value of total proteins in serum?*

   A. 6 - 8 g/ dl

5. *What are functions of plasma proteins?*

   A. Maintenance of plasma osmotic pressure, transport functions, defence function by immunoglobuliuns, coagulation, fibrinolysis, buffering of H^+. Plasma proteins have special; functions, including protease inhibitors e.g. α1-antitrypsin and α2-macroglobulin.

6. *Name some transport proteins.*

   A. Apolipoproteins (lipids), thyroxin binding globulin (thyroid hormones), retinol binding protein, transcortin (cortisol), haptoglobin (hemoglobin), transferrin (iron), and hemopexin (free heme).

7. *What is the clinical manifestation of Alpha-1-anti-trypsin deficiency?*

   A. Emphysema and chronic lung infections.

8. *What is the protein resposible of maintaing plasma osmotic pressur?*

   A. Albumin.

9. *What is the effect of hypoproteinemia?*

   A. Edema.

10. *Edema due to hypoproteinemia is seen in which conditions?*

    A. Liver cirrhosis (due to deficient synthesis of albumin), malnutrition (due to deficient absorption of proteins), and nephrotic syndrome (due to excess loss of proteins).

11. *In blood, albumin carries what substances?*

A. Free fatty acids, bilirubin, salicylate, and calcium.

12. *Is it safe to give salicylate to infants having hemolytic diseases?*

A. No because bilirubin and salicylate compete for binding to albumin, and so it is not safe to give salicylate to infants having hemolytic disease.

13. *What are the functions of albumin?*

A. It maintains colloidal osmotic pressure of plasma, and it transports non-esterified fatty acid and bilirubin.

14. *Albumin is synthesized in which organ?*

A. Liver.

15. *Where is gammaglobulins synthesised?*

A. By reticulo-endothelial system (spleen, lymph nodes).

16. *Polymorphism is exhibited by which proteins?*

A. Haptoglobin, transferrin, and ceruloplasmin.

17. *Albumin globulin ratio is reversed in which conditions?*

A. Cirrhosis, chronic infections, nephrotic syndrome, and multiple myeloma.

18. *Name acute phase proteins.*

A. C-reactive protein, ceruloplasmin, and haptoglobin.

19. *What is ceruloplasmin?*

A. It is a copper containing enzyme (ferroxidase) seen in blood. It is an acute phase protein.

20. *What is the clinical significance of ceruloplasmin?*

A. Ceruloplasmin level in blood is decreased in Wilson's hepatolenticular degeneration.

21. *What are the carrier proteins of copper?*

A. Ceruloplasmin (90%) and albumin (10%).

22. *Hemopexin carries what?*

A. Free heme.

23. *How hemophilia is manifested?*

A. Non-stopping hemorrhage after minor injuries.

24. *How hemophilia is transmitted?*

A. It is inherited as an x-linked recessive trait, males are affected, females are carriers.

25. *Hemorrhage tendency is seen in which conditions?*

A. Hemophilia, Vitamin K deficiency, and thrombocytopenia.

26. *What are anticoagulants?*

A. Citrate, , heparin, dicumarol, EDTA and defibrination of blood.

27. *What is natural blood anticoagulants?*

A. Heparin.

28. *What are plasma proteins?*

A. Pre-albumin, albumin and globulins (α-1, α-2, β- and γ-globulins).

29. *What are types of α-1 globulins?*

A. Prothrombin, retinol binding globulin, transcortin, vitamin D-binding globulin, α1-antitrypsin, α1-acid glycoprotein and α1-fetoprotein.

30. *What are types of α-2 globulins?*

A. Ceruloplasmin, haptoglobin, α2-macroglobulin and thyroxine-binding globulin.

31. *What are types of β- globulins?*

A. Plasminogen, transferrin, β2-microglobulin and c-reactive protein.

32. *What are types of γ- globulins?*

A. They include all antibodies which are IgG, IgA, IgM, IgD and IgE.

33. *What are nonprotein nitrogenous compounds of plasma?*

A. Urea, uric acid, creatinine, and amino acids.

34. *What is serum?*

A. serum is plasma without clotting factors.

35. *What is the composition of plasma?*

A. Water (90%) and solids (10%). They include, organic and inorganic matters.

36. *What are organic matters of plasma?*

A. Proteins, lipids (plasma lipoproteins), carbohydrate (glucose and other blood sugars), nonprotein nitrogenous compounds (urea, uric acid, creatinine, etc..), hormones, enzymes, ketone bodies).

37. *What are inorganic matters of plasma?*

A. Na^+, K^+, Ca^{2+}, Cl^- , carbon dioxide.

38. *What is transferrin?*

A. This is a plasma glycoprotein that is synthesized in the liver and each molecule can carry 2 atoms of iron in ferric state (Fe^{3+}).

39. *How much transferrin can carry iron?*

A. The capacity of transferrin to carry iron is up to 180-450 ug iron/dl. This is known as total iron binding capacity of transferrin (TIBC). As the plasma iron is 60-160 ug/dl, thus only 30% of the TIBC of transferrin is saturated.

40. *What is total iron binding capacity?*

A. maximum amount of iron that can be carried by transferrin per deciliter.

41. *What are physical properties of urine?*

A. These include volume, odor, color, aspect, specific gravity, pH and deposits.

42. *What is normal urine volume?*

A. 800 – 2000 ml/day.

43. *What are variations of urine volume?*

A. During sleep (at night), the amount of urine is about 1/2 the amount formed during the day. Polyuria is the urine excretion of more than 2000 ml/day. Oliguria is the excretion of less than 500 ml/day. Anuria is the excretion of less than 125 ml/day.

44. *What are causes of polyurea?*

A. *Physiological polyuria* as in high fluid intake, and high protein diet (the end product of protein metabolism is urea which causes osmotic diuresis). *Pathological polyuria* as in diabetes mellitus (glucose causes osmotic diuresis) and in diabetes insipidus (due to lack of antidiuretic hormone).

45. *What are causes of oliguria?*

A. *Physiological oliguria* as in low fluid intake and at hot weather due to excessive sweating. *Pathological oliguria* or anuria as in urinary obstruction (by stones or tumor), excessive vomiting and diarrhea or due to shock and hemorrhage.

46. *Why urine gets the odor of ammonia if left for a long peroid?*

A. Due to decomposition of urea by bacteria and release of ammonia.

47. *What is the cause of yellow color of urine?*

A. The color of urine is due to 2 pigments; urochrome and urobilin.

48. *What are variations of urine color?*

A. It may result from many metabolic products, drugs and foods, red color due to hematuria, and Yellow-brown due to jaundice.

49. *What is the cause of turbidity of urine?*

A. Normally, freshly voided urine is transparent. Turbid urine may be associated with the presence of abnormal constituents e.g. pus (pyuria), red cells (hematuria), chyle (chyluria) and crystals (calcium phosphate or urate).

50. *What is specific gravity of urine?*

A. It is the density of urine compared with distilled water which has a density of 1.00 (liquid density/water density).

51. *What is the other name of specific gravity of urine?*

A. Urine relative mass density.

52. *What is the normal specific gravity of urine?*

A. 1015-1025 (for urine collected over 24-hours).

53. *What does high specific gravity of urine mean?*

A. The higher the specific gravity, is the more the dissolved solids in urine e.g. urea, uric acid, sugars.

54. *What is the diagnostic importance of measuring specific gravity of urine?*

A. It indicates the concentrating power of the kidney.

55. *What are variations of urine specific gravity?*

A. Decreased in cases of diluted urine as in diabetes insipidus and increased in cases of concentrated urine as in diabetes mellitus.

56. *What is the normal pH of urine?*

A. Normally, urine pH is acidic (about 6).

57. *What is the cause of acidity of urine?*

A. Acidity of urine results from conversion of basic phosphate (Na_2HPO_3) into acid phosphate (NaH_2PO_3) in distal convoluted tubules of the kidney.

58. Why urine gets less acidic after meals?

A. Due to the formation of gastric HCl occurring after meals is associated with absorption of more bicarbonate. The later is then excreted in urine making it alkaline. This process is called *alkaline tide.*

59. *What are causes of high acidity of urine (pH below 6)?*

A. High protein diet, metabolic and respiratory acidosis, urinary tract infection by a type of bacteria called: E. coli.

60. *What are causes of less acidity of urine (pH above 6)?*

A. High citrus fruits and vegetables, administration of some alkalies as sodium bicarbonate, potassium depletion, as it leads to alkalosis.

61. *What are types of urine deposits?*

A. Pus cells, red cells, epithelial cells, parasites, ova as bilharzial ova, Casts, and crystals.

62. *What is clinical significance of presence of pus in urine?*

A. it indicates urinary tract infection.

63. *What are types of epithelial cells in urine ?*

A. Squamous epithelium is normally present in female urine. They are derived from skin while urine passing female genital tract. Columnar or transitional epithelium are derived from kidney, ureter or bladder due to a variety of causes e.g. infection.

64. *What are casts ?*

A. These are cylindrical structures formed basically from mucoprotein in the distal convoluted tubules. After formation, they become loose and go down the tubules into the urine.

65. *What are types of crystals in urine?*

A. The most common stones are urate, oxalate, and phosphate crystals.

66. *What are non-protein nitrogenous compounds in urine ?*

A. Urea, ammonia, creatinine, creatine, uric acid and amino acids.

67. *What is normal urine urea?*

A. 20 – 40 g/day.

68. *What is the main solute of urine?*

A. Urea.

69. *What are conditions of increased urea in urine?*

A. Increased protein intake or in conditions where protein catabolism is increased as in fever and diabetes mellitus.

70. *Where is ammonia derived from?*

A. Deamination of amino acids mainly glutamine by glutaminase enzyme.

71. *What are causes of ammonia variations in urine?*

A. In acidosis as in diabetes melitus, ammonia is increased. In alkalosis, ammonia is almost absent in urine.

72. *What does creatinine excretion depend on?*

A. It depends on ***muscle bulk*** of the individual and not on diet.

73. *Where is urine uric acid derived from?*

A. It is derived either exogenously from diet or endogenously from breakdown of tissue nucleoproteins in liver.

74. *Why uric acid is acidic?*

A. Because it contains 3 enol groups (C-OH) that can give (H^+) ions.

75. *Why uric acid sòlution is alkaline when we test it in the lab?*

A. Because uric acid is dissolved in an alkaline medium.

76. *What are conditions of increased uric acid in urine?*

A. Leukemia, severe liver disease and gout.

77. *What is allantion?*

A. It is a substance derived from partial oxidation of uric acid. It is present in birds urine. Human urine contains very small amount of allantion.

78. *What is amino aciduria?*

A. Excessive excretion of amino acids.

79. *What are causes of amino aciduria?*

A. In terminal liver disease due to failure of deamination of amino acids and in kidney disease due to inherited tubular defect in reabsorption of amino acids.

80. *What are abnormal constituents of urine?*

A. Sugars, proteins, ketone bodies, bilirubin and porphyrins.

81. *What are sugar excreted in urine?*

A. Glucose (glycosuria), fructose: (fructosuria), galactose (galactosuria) and pentose (pentosuria). Lactose (lactosuria) may occur in infants and in mothers during pregnancy, lactation and the weaning period.

82. *Are proteins normally excreted in urine?*

A. Normal urine contains very little amount of proteins (less than 30 mg/liter). These are albumin and globulins (30%) and mucoproteins of renal origin called Tamm Horsfall mucoprotein (70%).

83. *What is microalbuminuria?*

A. It is the excretion of proteins (30 – 200 mg/liter).

84. *What does microalbuminuria indicates?*

A. It indicates early affection of kidney as in diabetes mellitus.

85. *What is proteinuria?*

A. It is an increase of protein loss in urine (more than 200 mg/liter). It is a characterstic of all acute and chronic kidney diseases.

86. *What is the most common protein excreted in urine?*

A. Albumin (albuminuria) due to its low molecular weight and higher plasma concentration compared with globulins.

87. *What are other proteins excreted in urine?*

A. Bence Jones protein, hemoglobin and myoglobin.

88. *What is Bence Jones protein?*

A. It is an abnormal type of globulins (light chains of immunoglobulins), present in urine of patients suffering from multiple myeloma (malignant plasma cells).

89. *What are the characteristics of Bence Jones protein ?*

A. It is precipitated at $60^{\circ}C$, dissolved at $100^{\circ}C$ and reprecipitated on cooling.

90. *What does hemoglobinuria indicate?*

A. It indicates intravascular hemolysis as in malaria and hemolytic anemia.

91. *What does myoglobinuria indicate?*

A. It indicates massive muscle damage as in burns and severe electric shock.

92. *What are normal ketone bodies excreted in urine?*

A. Less than 18 mg.

93. *What is ketonuria?*

A. Ketonuria is the presence of ketone bodies in the urine in abnormal concentrations.

94. *What are causes of ketonuria?*

A. Ketonuria may occur in any condition where carbohydrate utilization is impaired e.g. starvation, carbohydrate poor diet and diabetes mellitus.

95. *What are causes of presence of bilirubin in the urine?*

A. Obstructive jaundice and in some stages of toxic jaundice.

96. *What is the color of jaundiced urine?*

A. Greenish brown color.

97. *What are causes of hematuria?*

A. It is caused by urinary bilharziasis, glomerulonephritis, traumatic or malignant diseases.

98. *What are normal porphyrins excreted in urine?*

A. Normally, 60 – 300 microgram coproporphyrins/day are excreted in urine.

99. *What are causes of the presence of excess amount of porphyrins in urine?*

A. Occurs in patients suffering from porphyrias.

100. *What are types of proteinuria?*

A. *Prerenal proteinuria* as in heart failure due to renal venous congestion. *Renal proteinuria* due to kidney affection (glomerular as in glomerulonephritis, or tubular as in pyelonephritis). *Postrenal proteinuria* due to lower urinary tract affection as inflammation or tumour of urinary bladder.

101. *What is milk?*

A. It is the secretion of mammary glands after labor.

102. *What is the major difference between human and cow's milk?*

A. Human milk has higher carbohydrate content than cow's milk while protein content is less.

103. *What is meant by humanization of milk?*

A. This is a process by which cow's milk is made to be as near as human milk, to suit the infant's needs.

104. *How is to humanize cow's milk?*

A. To humanize cow's milk, protein is to be half diluted and carbohydrate is to be added. Thus, to one cup of cow's milk, add half a cup of water and two tea-spoons of sugar. Iron, vitamin C and vitamin D may be added. This will make it comparable to human milk.

105. *What is the aim of humanization of milk?*

A. The aim of humanization is to decrease the concentration of casein which if present in high concentrations, forms a dense clot in the infant's stomach and leads to vomiting.

106. *How much is milk fat?*

A. 3.7 g/dl.

107. *Milk contains which type of fatty acids?*

A. The fatty acids are mainly unsaturated fatty acids (52%) and saturated (48%), half of them are medium chain saturated fatty acids (Lauric and Myristic acids).

108. *What is the advantage of medium chain fatty acids?*

A. They are easily digested, absorbed and metabolized.

109. *What are human milk proteins?*

A. Milk proteins: 1.2 g/dl. They are casein(25%), albumin, globulin (75%) and enzymes.

110. *What is the significance of albumin and globulins in human milk?*

A. Albumin and globulin (75%) are soluble i.e. easily digested. They also contain γ-globulins which give immunity for the baby.

111. *What is the major protein in cow's milk?*

A. 75% protein of cow's milk is casein.

112. *What type of protein is casein?*

A. It is a phosphoprotein of high biological value. It combines with calcium ions to form the insoluble calcium caseinate (milk clot).

113. *How the phosphate group is attached to protein?*

A. The phosphate groups are added to the hydroxyl groups of serine or threonine residues.

114. *What is milk clot?*

A. This is the enzymatic precipitation of milk casein by rennin enzyme.

115. *What is the significance of milk clot formation?*

A. This prevents the rapid passage of milk from the stomach to intestine and gives the sense of fullness.

116. *What are milk enzymes?*

A. Proteinase, amylase, peroxidase, catalase, alkaline phosphatase and aldehyde oxidase.

117. *Which contains more proteins, human or cow's milk?*

A. Milk proteins are less in human than in animal milk.

118. *What is whey?*

A. If milk is acidified and pH lowered to 4.7, the casein is precipitated (iso-electric precipitation). The supernatant is called whey. It contains proteins and all water soluble vitamins.

119. *What are the proteins present in whey?*

A. Lactalbumin, lactoglobulin and lysozyme.

120. *What is colostrum?*

A. It is the milk secretion in the first few days after labor.

121. *What is lactoferrin?*

A. It is iron binding protein present in milk, neutrophils and other body fluids. It has antibacterial action.

122. *What is skimmed milk?*

A. It is milk without milk fat.

123. *What is milk souring?*

A. It is the precipitation of casein by lowering milk pH to 4.6 (isoelectric point of casein). This is due to production of lactic acid from lactose by the action of bacteria. This is the idea of yoghurt formation.

124. *What is dried milk?*

A. Milk from which water is removed by lypholyzation.

125. *What is milk curdling?*

A. It is the precipitation of casein by milk clotting enzymes (rennin), in the form of insoluble calcium paracaseinate. This is the idea of cheese formation.

126. *What is milk pasteurization?*

A. It is sterilization of milk by heating it to 60°C for 30 minutes (or 70°C for 15 minutes) followed by rapid cooling.

127. *What is milk carbohydrate?*

A. lactose (7.0 g/dl).

128. *Which contains more lactose, human or cow's milk?*

A. Lactose is more in human than in animal milk.

129. *Why baby can consume large amounts of milk without developing nausea?*

A. Because the sweetness of milk is entirely due to lactose, which is less sweet than ordinary cane sugar; sucrose.

130. *What is the advantage of lactose over any other sugar?*

A. Lactose on hydrolysis gives glucose and galactose sugars. Glucose is a good source of energy. Galactose has a special value for the rapid synthesis of glyco- and galactolipids → Important for CNS.

131. *What are milk vitamins?*

A. Milk contains most of the vitamins. It is very rich in vitamins A and B_2. It is poor in vitamins C, D, and K.

132. *What are milk minerals?*

A. milk is the richest source of calcium and phosphorus. It is poor in iron sodium and potassium.

133. *What are physical properties of milk?*

A. Color is white due to the presence of fat globules and calcium phosphate. Cow's milk is creamy as it contains excess carotene. pH: 6 – 7.7. Specific gravity is 1032 at 32°C (human milk) and 1028 at 32°C (cow's milk).

134. *Which has higher specific gravity, full cream or skimmed milk?*

A. When milk is skimmed, the specific gravity rises (1033-1037) owing to the removal of fat (light constituent).

135. *Compare between colostrum and ordinary milk*

	Colostrum	Ordinary milk
Color	Yellowish as it contains excess carotene	White
Consistency	Thick	Thin
Reaction	Alkaline	Neutral
Proteins	15 g/dl and contains excess gamma globulins	1.2 g/dl
Carbohydrates	Less	7 g/dl
Lipids	Less	3.7 g/dl
Minerals	More	Less
Trypsin inhibitor	Present	Absent

136. *Compare between human and cow's milk*

	Human milk	Cow's milk
Proteins:	1.2 g/dl	3.3 g/dl
Casein	0.3 g/dl	2.7 g/dl
Albumin &glob.	0.9 g/dl	0.6 g/dl
Lactose	7.0 g/dl	4.7 g/dl
Lipids	3.7 g/dl	3.7 g/dl
Sat. FA	48%	58%
Unsat. FA	52%	42%
Minerals	Less	More
Vitamins	More	Less

137. *Is milk a complete diet?*

A. Milk is considered a complete diet because it contains almost all substances necessary to maintain life (carbohydrate, proteins, lipids, minerals and vitamins). However, it is not a perfect diet because It is deficient in vitamins: C, D and K iron and copper.

MCQ, Matching, true and false and Completion

Select and encircle the most appropriate answer or completion:

1. *Human milk:*
   A. Contains saturated fatty acids more than unsaturated fatty acids.
   B. Poor in vitamin A.
   C. Contains more lactose than cow's milk..
   D. Contains more proteins than cow's milk.
   E. Needs supplementation of calcium

2. *Which statement about colostrum is INCORRECT:*
   A. It is poor in proteins especially gamma globulins.
   B. It is yellow in color due to the presence of excess carotene.
   C. It is the milk secreted immediately after birth.
   D. It contains less fat and less carbohydrate than late milk

3. *Ferritin:*
   A. Is a plasma protein which binds iron
   B. Is a muscle protein which oxidizes iron to the ferric state
   C. Is a protein which stores ferric ions
   D. Is involved in absorption of vitamin B_{12} from intestine
   E. Regulates excretion of urinary iron

4. *Polyurea is:*
   A. A sense of thirst associated with diabetes mellitus.
   B. Excessive urea formation.
   C. Decreased daily urine volume below normal range.
   D. Increased daily urine volume above normal range.

5. *In jaundice, the color of the urine is:*
   A. Amber yellow
   B. Greenish brown
   C. Red
   D. Purple

6. *Specific gravity of 24 hours urine is ranged between:*
   A. 1000-1010
   B. 1005-1010
   C. 1015-1025
   D. 1040-1050

7. *The acidity of the urine is due to:*
   A. Conversion of basic phosphate into acid phosphate.
   B. HCl formation after meal.
   C. Action of glutaminase on glutamine with release of ammonia.
   D. Excretion of uric acid in urine.

8. *All the followings are constituents of normal urine EXCEPT:*

A. Uric acid.
B. Bence-Jones protein.
C. Urea.
D. Creatinine.

9. *The major source of ammonia in the kidneys is:*

A. Leucine
B. Glutamate
C. Glutamine
D. Aspartate
E. Asparagine

10. *After meal, alkaline tide of urine is due to:*

A. Excess lipids in diet.
B. Excess carbohydrates in diet.
C. Excess proteins in diet.
D. Gastric HCl formation and excretion of excess bicarbonate.

11. *One of the non protein nitrogenous compounds in urine is:*

A. *Gluguronic acid*
B. Uric acid
C. Carnitine
D. Urobilinogen

12. *Creatinine in urine is derived from:*

A. Liver.
B. Skeletal muscles.
C. Spleen.
D. Protein of diet.

13. *Lactose is found in urine in case of:*

A. Diabetes insipidus.
B. Lactation.
C. Muscle diseases.
D. Acidosis.

14. *The followings are abnormal constituents of urine EXCEPT:*

A. Bilirubin.
B. Ketone bodies.
C. Pentoses.
D. Indican.

15. *The main solute of urine is:*

A. Sodium chloride.
B. Urea.
C. Uric acid.
D. Ammonia.

16. *Casein of milk is:*

A. Derived protein.
B. Glycoprotein.
C. Phosphoprotein.
D. Lipoprotein.

17. *Insoluble calcium caseinate is the milk clot that:*

A. Makes the milk pH slightly alkaline.
B. Contains excess antibodies.
C. Gives infant the sense of fullness.
D. Gives the sweetness of the milk.

18. *Milk is the richest source of:*

A. Iron.
B. Calcium.
C. Copper.
D. Zinc.

19. *Among the anti-coagulants normally present in blood is:*

A. EDTA.
B. Dicumarol.
C. Sodium citrate.
D. Heparin.

20. *A substance that can prevent blood clotting is:*

A. Sodium citrate.
B. Sodium chloride.
C. Protamine sulfate.
D. Fibrinogen.

21. *The normal pH of the blood is:*

A. 6.8.
B. 7.4.
C. 7.7.
D. 8.0

22. *Which finding about iron deficiency anemia is INCORRECT:*

A. Plasma iron is decreased.
B. Plasma TIBC is decreased.
C. Plasma ferritin is decreased.
D. RBCs are hypochromic and microcytic.

23. *Blood plasma differs from blood serum in content of:*

A. Lipids.
B. Carbohydrates.
C. Proteins.
D. Non-protein nitrogenous compounds.

24. *Which of the following is not anticoagulant:*

A. EDTA.
B. Vitamin K.
C. Potassium oxalate.
D. Heparin.

25. *Which statement about seminal fluid is INCORRECT:*

A. Glucose is the main carbohydrate.
B. Zinc is very important constituent.
C. Sperms are formed of head, neck and tail.
D. Lipid content is rich in prostaglandins.

26. *The followings are functions of CSF except:*

A. It protects the brain and spinal cord during movement.
B. It minimizes the friction between vertebrae during movement.
C. It carries nutrients to brain and spinal cord.
D. It maintains constant pressure inside the head and the spinal cord.

Answer Key

MCQ:

1	2	3	4	5	6	7	8	9	10
C	A	C	D	B	C	A	B	C	D
11	12	13	14	15	16	17	18	19	20
B	B	B	D	B	C	C	B	D	A
21	22	23	24	25	26				
B	B	C	B	A	B				

Chapter 21 Acid Base Balance and pH

1. *Relationship between pH and pK is given by which equation?*

   A. Henderson-Hasselbalch's equation.

2. *What determines the pH of buffer?*

   A. By the ratio of salt to acid.

3. *Buffer is most effective when?*

   A. When pK of the acid is nearer to pH.

4. *When is the buffering capacity is more?*

   A. When the absolute concentrations of salt and acid are more.

5. *In the blood, which buffer is most effective?*

   A. Bicarbonate buffer.

6. *What are the mechanisms for maintaining the normal pH of plasma?*

   A. Buffers of plasma, lung mechanism, and kidney mechanism.

7. *What is the alkali reserve of the body?*

   A. Bicarbonate is the alkali reserve.

8. *What is the ratio of bicarbonate to carbonic acid in blood?*

   A. Bicarbonate to carbonic acid ratio is 20.

9. *What are the mechanisms by which renal regulation of acid load is achieved?*

   A. Excretion of hydrogen ions in urine, excretion of ammonium ions in urine, and production of bicarbonate in renal tubules.

10. *Glutaminase enzyme is used for what purpose?*

    A. For production of ammonia in kidney tubules.

11. *What is metabolic acidosis?*

    A. Primary deficit of bicarbonate.

12. *What are the causes of metabolic acidosis?*

    A. Diabetic ketosis, chronic renal failure, and diarrhea.

13. *What are the features of diabetic ketoacidosis?*

    A. Lowered bicarbonate, elevated plasma chloride, and increased anion gap.

14. *What is the formula used to calculate anion gap?*

A. (Sodium + potassium) minus (chloride + bicarbonate).

15. *What is the cause for high anion gap acidosis?*

A. Diabetic ketoacidosis, chronic renal failure, renal tubular acidosis, lactic acidosis.

16. *What is metabolic alkalosis?*

A. Primary excess of bicarbonate.

17. *What are the causes of metabolic alkalosis?*

A. Prolonged vomiting, gastric aspiration, and ingestion of antiacids.

18. *What is respiratory acidosis?*

A. Primary excess of carbonic acid.

19. *What are the causes of respiratory acidosis?*

A. Bronchial asthma, bronchopneumonia, narcotic poisoning and emphysema.

20. *What is respiratory alkalosis?*

A. Primary deficit of carbonic acid.

21. *What is the cause for respiratory alkalosis?*

A. Hyperventilation.

22. *What are the results of prolonged vomiting?*

A. Alkalosis, hypochloremia, and hypokalemia.

23. What is the pH of 0.1 M hydrochloric acid?

A. One.

24. *When pH falls by 1 unit, what is the change in the hydrogen ion concentration?*

A. Increases by 10 times.

MCQ, Matching, True and False and Completion

Select and encircle the most appropriate answer or completion:

1. *Respiratory acidosis may be caused by:*
   A. Hyperventillation
   B. Emphysema
   C. Vomiting
   D. Ingestion of excessive amounts of sodium bicarbonate
   E. Starvation

2. *Which of the following is likely to produce metabolic acidosis?*
   A. High altitude
   B. Excessive vomiting
   C. Starvation
   D. Toxic dose of morphine
   E. Ingestion of excessive amounts of sodium bicarbonate

3. *The pH of a solution is equal to:*
   A. The hydrogen ion concentration(H^+)
   B. $Log_{10}(H^+)$
   C. $- log_{10}(H^+)$
   D. $ln(H^+)$
   E. $- ln (H^+)$

4. *The ionization of a weak acid HA ⇆ H+ + A- has an apparent equilibrium constant, K, which is equal to:*
   A. Log $[A^-]/[HA]$.
   B. $[HA]/[A^-]$.
   C. $[H^+][A^-]/[HA.]$
   D. $[A^-]/[HA]$.
   E. $[HA]/[H^+][A^-]$.

5. *When the pH for a solution of the acid described in the previous question is equal to the pK, the ratio of the concentrations of the salt and the acid $[A^-]/[HA]$:*
   A. 0
   B. 1
   C. 2
   D. 3
   E. 4

6. *All the following are causes of respiratory alkalosis <u>EXCEPT</u>:*
   A. Hysteria
   B. Fevers
   C. Early stage of salicylate poisioning
   D. Hepatic failure
   E. Mechanical asphyxia

7. *In respiratory alkalosis:*
   A. The acute stage is associated with an abnormally low plasma [HCO_3^-]
   B. The mechanism of compensation causes an increase in the plasma [HCO_3^-]
   C. The plasma pH never returns to the normal range in the fully compensated state
   D. In the partially compensated state there will be a negative base excess equal to the difference between 24 mEq/L and the actual plasma [HCO_3^-]
   E. Compensation involves changing P_{CO_2}

In the following questions indicate with clear (T) the true statements, and with clear (F) the false statements:

The following statements are correct:

8. An acid is a substance which can dissociate to produce hydrogen ions.
9. A base is a substance which can accept hydroxyl ions.
10. Buffering is the process by which a strong acid is replaced by a base.
11. pH is the $\log_{10}$ of the hydrogen ion concentration.
12. pH of the blood is 7.

Renal mechanism in the handling of bicarbonate include:

13. Passage through the glomeruli by filtration.
14. Absorption as bicarbonate from the tubular lumen.
15. Dissociation into sodium and bicarbonate ions under the influence of carbonate dehydratase.
16. Diffusion of bicarbonate ion from tubular cells to plasma down a concentration gradient.
17. Active secretion of hydrogen ions from tubular cells into the tubular lumen.

Ammonium ions:

18. Forms a buffer pair with phosphate.
19. Diffuses out of tubular cells into the urine much more rapidly than ammonia.
20. Dissociates in the tubular cell to liberate hydrogen ions.
21. Produced in the tubular cell leads to increased renal excretion of bicarbonate.
22. Is formed from glutamine, together with glutamate which is ultimately converted to glucose.

Important buffering activity is provided by:

23. Bicarbonate in blood.
24. Hemoglobin.
25. Phosphate in the blood.
26. Phosphate in urine.
27. Plasma proteins.

In metabolic alkalosis:

28. There is marked hypoventilation.
29. The primary abnormality is a rise in plasma bicarbonate.
30. Tetany may develop in spite of normal total plasma calcium levels.
31. Metabolic compensation depends on increased urinary bicarbonate loss.
32. With chloride depletion, the urine may be inappropriately acidic.

***Matching:** For each set of numbered questions, choose the **ONE BEST** answer from the list of lettered options below it. An answer may be used once or more times, or not at all.*

33. *Proton donor*
34. *Proton acceptor*
35. *Buffer*
36. *Pk*
37. *pH*

A. Negative logarithm of hydrogen ion concentration.
B. Is a solution, which resists the change in pH
C. A base
D. An acid
E. Acid dissociation constant

38. *Respiratory acidosis*
39. *Metabolic acidosis*
40. *Respiratory alkalosis*
41. *Metabolic alkalosis*

A. Is due to primary decrease in the concentration of carbonic acid content (CO_2 tension).
B. Is due to a primary increase in the concentration of carbonic acid content (CO_2 tension).
C. Is due to a primary increase in the concentration of bicarbonate ($NaHCO_3$).
D. Is due to a primary decrease in the concentration of bicarbonate ($NaHCO_3$).

Answer Key

MCQ:

1	2	3	4	5	6	7
B	C	C	C	B	E	A

True and false:

8	9	10	11	12	13	14	15	16	17
T	F	F	F	F	T	F	F	T	T
18	19	20	21	22	23	24	25	26	27
F	F	T	F	T	T	T	F	T	F
28	29	30	31	32					
F	T	T	T	T					

Matching:

33	34	35	36	37	38	39	40	41
D	C	B	E	A	B	D	A	C

Chapter 22

Hormones and Mechanism of Action of Hormones

1. *What are G proteins?*

   A. They are involved in signal transduction.

2. *How the G proteins work?*

   A. The receptors are in membrane, and binding of hormone causes activation of G protein.

3. *How is G protein activated?*

   A. Binding of hormone on receptor causes attachment of GTP to G protein, and thereby G protein is activated.

4. *What is the function of activated G protein?*

   A. Activated G protein activates adenylate cyclase enzyme.

5. *How adenylate cyclase is destroyed?*

   A. By phosphodiesterase enzyme.

6. *How cyclic AMP further works?*

   A. Cyclic AMP activates protein kinase.

7. *What is the function of protein kinase?*

   A. It phosphorylates enzymes or target proteins, so that they are activated.

8. *Name some hormones, which act through cyclic AMP as the second messenger.*

   A. Glucagon, ACTH, TSH, ADH, FSH, and LH.

9. *Name some second messengers.*

   A. Cyclic AMP, 1,2-diacylglycerol, inositol triphosphate, and calcium.

10. *Name some hormones, which act through cyclic GMP as the second messenger.*

    A. ANF (atrial natriuretic factor).

11. *Name some hormones that bind to intracellular receptors.*

    A. Glucocorticoids, mineralocorticoids, estrogens, progesterone, androgens, and thyroxin.

12. *What are hypothalamic neuropeptides?*

A. These neurohormones are antidiuretic hormone (ADH) and oxytocin.

13. *Name the important hypothalamic releasing factors.*

A. TRH (thyrotropin releasing hormone), GnRH (gonadotrophin releasing hormone), GHRH (growth hormone releasing hormone), somatostatin (growth hormone inhibitory factor), CRF (corticotrophin releasing factor), and PIF (prolactin inhibitor factor).

14. *α-Chain of human chorionic gonadotrophin (HCG) is shared with what else?*

A. TSH, FSH, LH and HCG have the common α chain, and β chains are specific.

15. *What are the hormones produced by anterior pitutary?*

A. GH (Growth hormone), ACTH (Adrenocorticotropic hormone), LH (Luteinizing hormone), FSH (Follicle stimulating hormone), TSH (Thyroid stimulating hormone), MSH (Melanocyte stimulating hormone), and PRL (Prolactin).

16. *What is the major effect of growth hormone(GH)?*

A. GH increases the uptake of amino acids by cells, enhances protein synthesis, and produces positive nitrogen balance. The anti-insulin effect of GH causes lipolysis and hyperglycemia. The overall effect of GH is to stimulate growth of soft tissues, cartilage, and bone. It is anabolic.

17. *Increased secretion of growth hormone will lead to what condition?*

A. Excess secretion of GH secretion leads to gigantism in children and acromegaly in adults.

18. *What is the result of decreased growth hormone secretion?*

A. Deficiency of GH secretion in early childhood results in pituitary dwarfism.

19. *What is the function of FSH?*

A. FSH stimulates growth of ovarian follicles in females and spermatogenesis (Sertoli cells) in males.

20. *What is the function of LH?*

A. Testosterone in males (secreted by Leydig interstatial cells) and progesterone in females (secreted by corpus luteum), are increased under the influence of LH.

21. *The 11-hydroxylase is required for the synthesis of which hormone?*

A. Cortisol.

22. *The 17-hydroxylase is required for the synthesis of which hormones?*

A. Cortisol, testosterone, and estradiol.

23. *Which is the precursor of steroid hormones?*

A. Cholesterol.

24. *What are the hormones produced from progesterone?*

A. Corticosterone, aldosterone, testosterone, and estrogens.

25. *Cortisol has how many carbon atoms?*

A. 21.

26. *Aldosterone has how many carbon atoms?*

A. 21.

27. *Testosterone has how many carbon atoms?*

A. 19.

28. *Estrogen has how many carbon atoms?*

A. 18.

29. *What is the immediate precursor of estrogens?*

A. Testosterone.

30. *What are the structural features of estradiol?*

A. Cyclopentanophenanthrene ring, total 18 carbon atoms, aromatic character of A ring, and hydroxyl groups on 3rd and 17 carbon atoms.

31. *21-hydroxylase is required for the synthesis of which hormone?*

A. Aldosterone.

32. *What are the effects of glucocorticoids?*

A. Increased gluconeogenesis, and lipolysis, elevated protein breakdown, and depressed immune function.

33. *What is the precursor of thyroxin?*

A. Tyrosine.

34. *Name some antithyroid agents.*

A. Thiocyanate, perchlorate, and methimazole.

35. *How thyroid hormones are produced?*

A. Tyrosine residues of thyroglobulin are iodinated.

36. *What is the function of thyroid stimulating hormone?*

A. It increases the uptake of iodine by thyroid gland, enhances the oxidation of iodine to iodide, and favors the hydrolysis of thyroglobulin to produce T4.

37. *What is the ratio of T4 and T3 in blood?*

A. Blood concentration of T4 is 70 times more than T3.

38. *What are the functions of thyroid hormones?*

A. Calorigenic effect or thermogenesis and BMR is increased.

39. *How this is produced?*

A. The thermogenic effect is mediated by uncoupling of oxidative phosphorylation. Thyroxin in large quantities can swell the mitochondria. Basal metabolic rate (BMR) is increased.

40. *What are the biochemical features of thyroid hormones?*

A. Thyroxin increases cellular metabolism. Gluconeogenesis and carbohydrate oxidation are increased. Glucose tolerance test shows rapid absorption. Fatty acid metabolism is increased.

41. *Deficiency of thyroxine results in which condition?*

A. Myxedema.

42. *What are the salient features of hypothyroidism?*

A. Decreased T3 level, increased TSH level, lethargy, hypercholesterolemia, weight gain, and decreased basal metabolic rate.

43. *What are the characteristic features of primary hyperthyroidism?*

A. High TSH and T4 levels, increased rate of metabolism, weight loss, tachycardia, fine tremors, sweating, diarrhea, emotional disturbances, anxiety, and sensitivity to heat.

MCQ, Matching, True and False and Completion

Select and encircle the most appropriate answer or completion:

1. *The parathyroid gland regulates the metabolism of:*
   A. Calcium
   B. Phosphate
   C. Both of the above (A and B)
   D. Magnesium and phosphate
   E. Sodium and phosphate

2. *A hyperglycemic factor produced by the pancreas is:*
   A. Insulin
   B. Lipase
   C. Glucagon
   D. FSH
   E. Thyroxin

3. *A substance present in the small intestine that stimulates contraction of the gall bladder is:*
   A. Rennin
   B. Secretin
   C. Cholecystokinin
   D. Enteropeptidase
   E. Gastrin

4. *Thyroxin is synthesized in the thyroid gland from:*
   A. Tryptophan
   B. Indole
   C. Histidine
   D. Tyramin
   E. Thyroglobulin

5. *In insulin deficiency:*
   A. Protein synthesis is depressed
   B. Protein degradation is increased
   C. Nitrogen excretion is increased
   D. Fatty acid synthesis is depressed
   E. All of these

6. *The prostaglandins:*
   A. Cause hypertension
   B. Occur only in prostatic tissue
   C. Synthesis needs cyclo-oxygenase
   D. Are synthesized from oleic acid
   E. Have chemotactic action

7. *Gigantism and acromegaly are diseases due to alterations in secretion of:*
   A. FSH
   B. TSH
   C. Growth hormone
   D. Parathyroid hormone
   E. Melatonin

8. *In mammals, norepinephrine is synthesized from:*
   A. Epinephrine
   B. Adrenaline
   C. Indole
   D. Tyrosine
   E. Serotonin

9. *Progesterone is a precursor of:*
   A. Aldosterone, cortisone, and corticosterone
   B. Cholesterol, cortisone, and cholic acid
   C. Aldosterone, deoxycholic acid, and pregnenolone
   D. Hydrocortisone, pregnenolone, and esteriol
   E. Corticosterone, testosterone, and cholesterol sulfate

10. *Two actions of insulin are:*
   A. Promotes the conversion of fat to carbohydrate and facilitates the passage of glucose into the cells
   B. Facilitates the passage of glucose into the cells and decreases fatty acid oxidation.
   C. Stimulate hexokinase and promotes gluconeogenesis
   D. Promotes gluconeogenesis and inhibits lipolysis
   E. Inhibits lipolysis and promotes glycogenolysis

11. *Glucagon and epinephrine are similar in that:*
   A. Both promote glycogenolysis in liver and muscle
   B. Both promote the activation of liver phosphorylase
   C. Both reduce blood glucose level
   D. Both are formed by the α–cells of the pancreas
   E. Both are hypertensive agents

12. In hypoparathyroidism in man there occurs:
   A. Elevation of serum calcium levels
   B. Urinary calculi
   C. Increase of calcium absorption
   D. Decrease in ionic serum calcium
   E. Decrease in serum thyroxin

13. *What serves as a precursor of testosterone?*
   A. Andrenosterone
   B. Pregnenolone
   C. Estrone
   D. Methyltestosterone
   E. Aldosterone

14. *Chemically, the steroids are derivatives of:*
   A. Fatty acids
   B. Cholesterol
   C. Ergosterol
   D. Perhydrocyclopentanophenantherene
   E. None of these

15. *Measurement of urinary 17-ketosteroids:*
   A. Gives abnormally high values in pheochromocytoma
   B. Gives abnormally high values in children
   C. Used for evaluating adrenal and testicular function
   D. Is an indicator of functioning adrenal glands
   E. Gives abnormally high values in hypogonadism

16. *Which of the following hormones or conditions DOES NOT cause increased lipolysis?*
   A. Insulin
   B. Glucagon
   C. Epinephrine
   D. Diabetes mellitus
   E. Starvation

17. *Pairs of hormones with antagonistic effects include all of the followings EXCEPT:*
   A. Vasopressin – oxytocin
   B. Insulin – glucagon
   C. Calcitonine – parathyroid hormone
   D. Melanocyte stimulating hormone – melatonin
   E. Histamine – serotonin

18. *Which of the following activities is NOT increased following exposure to physiologic concentrations of insulin?*
   A. Plasma membrane transfer of glucose
   B. Glucose oxidation
   C. Gluconeogenesis
   D. Lipogenesis
   E. Glycogenesis

In the following questions indicate with clear (T) the true statements, and with clear (F) the false statements:

In diabetes;

19. Glucose -6- phosphatase activity increases
20. Glucose -6- phosphate dehydrogenase activity increases
21. Glucose utilization is impaired
22. Gluconeogenesis is impaired
23. Glycogen synthase activity increases

Progesterone:

24. Differs from deoxycorticosterone at C-21
25. Can be synthesized by the ovaries and adrenals
26. Is excreted in the urine as pregnanediol glucosiduronate
27. Is relatively inactive when given orally
28. Stimulates breast acini

Glucagon effects include:

29. Activation of phosphorylase kinase
30. Inhibiting gluconeogenesis
31. Causing glycogenolysis
32. Activating muscle phosphorylase
33. Inhibiting lipolysis

***Matching:** For each set of numbered questions, choose the **ONE BEST** answer from the list of lettered options below it. An answer may be used once or more times, or not at all.*

34. *A polypeptide secreted by pancreatic α-cells, that increases blood sugar levels*
35. *Increases insulin release from the pancreas*
36. *Enhance protein synthesis*
37. *Causes release of calcium from bone*
38. *Increases glycogenolysis in liver and muscles*

A. Sulfonylurea
B. Epinephrine
C. Thyroxine
D. Glucagon
E. Parathyroid hormone

39. *Contraction of uterine smooth muscle*
40. *Secretion of pepsinogen*
41. *Inhibits calcium release from bones*
42. *Stimulation of gluconeogenesis*
43. *Development of alveolar system of mammary glands*

A. Cortisol
B. Oxytocin
C. Progesterone
D. Gastrin
E. Calcitonin

44. *Immediate precursor of norepinephrine*
45. *Required for conversion of norepinephrine to epinephrine*
46. *End product of epinephrine and nor epinephrine catabolism*
47. *Produced by action of catechol O-methyl transferase (COMT) on epinephrine and norepinephrine*
48. *Formation is stimulated by epinephrine*

A. Dopamine
B. Normetanephrine and metanephrine
C. S-Adenosylmethionine
D. Cyclic AMP
E. Vanilylmandelic acid

49. *Mobilizes triacylglycerols*
50. *Releases milk*
51. *Antidiuretic hormone*
52. *Produced by the placenta*
53. *Stimulate testosterone production*
    A. Luteinizing hormone
    B. Vasopressin
    C. Glucagon
    D. Chorionic gonadotropin
    E. Oxytocin

54. Mammary gland
55. Uterus
56. Gall bladder
57. Skeleton
58. Smooth muscle
    A. Calcitonin
    B. Prostaglandins
    C. Oxytocin
    D. Progesterone
    E. Pancreozymin

Answer Key

MCQ:

1	2	3	4	5	6	7	8	9	10
A	C	C	E	E	C	C	D	A	B
11	12	13	14	15	16	17	18		
B	D	B	D	C	A	A	C		

True and false:

19	20	21	22	23	24	25	26	27	28
T	F	T	F	F	T	T	T	F	T
29	30	31	32	33					
T	F	T	F	F					

Matching:

34	35	36	37	38	39	40	41	42	43
D	A	C	E	B	B	D	E	A	C
44	45	46	47	48	49	50	51	52	53
A	C	E	B	D	C	E	B	D	A
54	55	56	57	58					
C	D	E	A	B					

Chapter 23

Cancer, Oncogenes, and Tumor Markers

1. *What are the most common causes of malignancy?*
   A. About 80% of human cancers are caused by environmental factors (pollutions), principally chemicals.
2. *Can life style cause cancer?*
   A. Yes. e.g. by cigarette smoking.
3. *Name some chemical carcinogens.*
   A. Methyl cholanthrene, aflatoxins, benzopyrenes and asbestos.
4. *How can alphatoxins be produced?*
   A. They are produced by the mold aspergillus flavus that may be present in some food.
5. *Name some physical carcinogens.*
   A. X-ray, gamma-ray and UV-ray.
6. *Name some anti-mutagens and anti-carcinogens.*
   A. Vitamin A, Vitamin C, Vitamin E and curcumin.
7. *Name some oncogenic viruses.*
   A. Epstein Barr virus (EBV), hepatitis B virus (HBV), and human papilloma virus.
8. *What are proto-oncogenes?*
   A. These are normal genes (about 100) present in normal cells in human genome. They have specific functions in cell growth and differentiation.
9. *What are Oncogenes?*
   A. Genes capable of causing cancer. Oncogenes are specific sequences in DNA which when expressed may produce cancer.
10. *Name a cancer, produced by the deletion of an onco-suppressor gene.*
    A. Retinoblastoma.
11. *What are properties of cancer cells?*
    A. They diminish control of growth, Invade local tissues, and spread (metastasis) to the other parts of the body.

12. *What are properties of benign cells?*

A. They diminish control of growth, do not invade local tissues, and do not spread to the other parts of the body.

13. *What are tumor suppressor genes?*

A. These genes suppress cancer (tumor) formation. Their protein product inhibits mitosis.

14. *What is retinoblastoma gene?*

A. Retinoblastoma is a cancerous tumor of the retina. It occurs in two forms, familial retinoblastoma which is multiple tumors in the retinas of both eyes occurring in the first weeks of infancy and sporadic retinoblastoma which is single tumor appears in one eye sometimes in early childhood before the retina is fully developed.

15. *What are causes of familial retinoblastoma gene?*

A. Occurs when the fetus inherits from one of its parents a chromosome (number 13) that contains a deleted or mutated RB gene. Then later in life, mutation of the remaining RB gene (somatic mutation) will remove the inhibition provided by RB protein ($p110^{RB1}$) → Retinoblastoma.

16. *What are causes of sporadic retinoblastoma gene?*

A. In this disease, both inherited RB genes are normal but later in life, both genes undergo somatic mutation (often a deletion) → Retinoblastoma.

17. *What is P^{53} gene?*

A. It is a tumor suppressor gene.

18. *Why it is called 53?*

A. Because its protein product is 53 kilo Daltons hence the name.

19. *What is the site of P^{53} gene?*

A. Short arm of chromosome 17 in somatic and gametogenic cells.

20. *What is the function of P^{53} gene?*

A. It prevents the formation of some tumors. Its mutation may lead to cancer lung, cancer breast and cancer colon.

21. *What is the mechanism of action of P^{53} gene?*

A. Regulation of cell division, control of DNA damage and repair, protection against viral infection, and P^{53} has a role in apoptosis.

22. *What is apoptosis?*

A. is a programmed cell death controlled by specific gene. Stimulation of this gene → Rapid death of the cells p^{53} participate and stimulate apoptosis by unknown mechanism.

23. *What are mechanisms of transformation of protooncogenes into oncogenes?*

A. This can be done by: promotor insertion(viral infection), enhancer insertion, chromosomal translocations and gene amplification.

24. *How can promoter insertion help transformation of protooncogenes into oncogenes?*

A. When certain viruses infect cells, a viral DNA copy (cDNA) is synthesized by reverse transcriptase and this cDNA is integrated in the host genome. These inserted new cDNA act as promoter of transcription of prooncogene → oncogene.

25. *How can promoter enhancer insertion help transformation of protooncogenes into oncogenes?*

A. Here the viruses that infect cells produce cDNA that act as enhancer (stimulator) which stimulate DNA for transcription.

26. *How can promoter chromosomal translocations help transformation of protooncogenes into oncogenes?*

A. A piece of one chromosome is split off and then joined to another chromosome. If the second chromosome donates material to the first, the translocation is said to be reciprocal. Example of chromosomal translocation include Philadelphia chromosome (involving chromosomes 9 and 22) → chromic granulocyctic leukemia. Chromosomal translocation of chromosomes 8 and 14 → cancer of human B lymphocytes → Burkitt's lymphoma.

27. *How can promoter gene amplification help transformation of protooncogenes into oncogenes?*

A. This is an increase the number of copies of normal proto-oncogene within the cell e.g. genes for certain enzymes e.g. dihydrofolate reductase. This results in an increase of enzyme activity → Resistance to certain anti-malignant drugs as methotrexate. Amplification may play a role in the progression of tumor cells to a more malignant state.

28. *How can promoter single point mutation help transformation of protooncogenes into oncogenes?*

A. Sometimes a single point mutation of proto-oncogene from normal human cells leads to production of mutant protein → cancer e.g. c-ras proto-oncogene from normal human cells and c-ras oncogene from a cancer of human bladder showed that they differed only in one base → an amino acid substitution at position 12 of certain protein that affect G-protein in cell membrane → affect adenylate cyclase → affect hormonal action.

29. *What are mechanisms of action of oncogenes?*

A. They may act on key intracellular pathways involved in growth control e.g. single point mutation may result in a protein that affect mitosis. The product of oncogenes may imitate the action of a polypeptide growth factor. The product of oncogene may also imitate an occupied receptor for growth factor.

30. *What are functions of proto-oncogenes?*

A. (1) Synthesis of protein kinase enzyme. This enzyme helps phosphorylation of some proteins changing its activity. (2) Synthesis of growth factors and growth factor receptors. (3) Some proto-oncogenes (c-Jun proto-oncogenes) act as regulator genes that regulate gene expression. (4) Some proto-oncogenes (c-ras-proto-oncogenes) code for synthesis of protein that binds GTP. GTP acts as first messenger for some hormones.

31. *What are growth factors?*

A. Growth factors are polypeptides exert a mitogenic response on their target cells. They affect many different types of cells e.g. blood cells, nervous system, mesenchymal tissues and epithelial tissues.

32. *How do growth factors act in endocrine, paracrine or autocrine manner ?*

A. Trey act as endocrine manner like hormones, they may be synthesized elsewhere in the body and pass in circulation to their target cells. Paracrine manner: they may be synthesized in certain cells and secreted from them to affect neighboring cells. However the cells that synthesize the growth factors are not themselves affected, because they lack suitable receptors. Autocrine manner: cells that synthesize growth factors have receptors for them or they are secreted inside cells and directly stimulate various processes.

33. *What is mode of action of growth factors?*

A. Growth factors bind with specific cell receptors on the plasma membrane of target cells forming growth factor receptor complex. This causes phosphorylation of target proteins in the cytoplasm. The growth factor- receptor complexes are subjected to endocytosis in coated vesicle (like LDL) → rapid activation of certain cellular proto-oncogene → Oncogenes.

34. *What are tumor markers?*

A. They are factors, synthesized and released by cancer cells or produced by the host cells in response to the presence of cancerous tissue. They could be detected in blood and therefore indicate the presence of the tumor in the body.

35. *What is the clinical application of tumor markers?*

A. They are useful for the following purposes. (1) For follow up of cancer and (2) to monitor the effectiveness of the therapy. (3) To detect the recurrence of the tumor.

36. *Where is tumor markers present?*

A. Tumor markers may be present in circulation, in body fluids or associated with cells In the cytoplasm or on cell membrane.

37. *What is the structure of tumor markers?*

A. Tumor markers may be enzymes, hormones, and proteins (tumor antigen).

38. *What is the ideal tumor marker?*

A. Ideally, tumor marker should provide the following uses in patients having cancer (1) Screening the asymptomatic population (2) Diagnosis the symptomatic patients. (3) Staging the disease. (4) Monitoring the response of the therapy. (5) Assessing prognosis. (6) Detecting recurrence.

39. *What are properties of ideal tumor marker?*

A. Have high disease sensitivity, high disease specificity, organ specific, its level reflects the stage of the disease, and must be stable.

40. *What does sensitivity mean?*

A. It should be positive in all patients with particular cancer.

41. *What does specificity mean?*

A. It should be negative in all normal population.

42. *What does stable mean?*

A. It is not subjected to marked fluctuation in stable disease state.

43. *Name some important tumor markers.*

A. α-Feto protein, CEA (carcino embryonic antigen), PSA (Prostatic specific antigen) and HCG (Human chorionic gonadotropin)

44. *Alpha feto protein (AFP) level in serum is increased in which condition?*

A. Hepatoma.

45. *Carcino embryonic antigen (CEA) level is increased in which type of cancers?*

A. Colorectal and gastrointestinal cancers.

46. *What is the significance of beta chain of human chorionic gonadotrophin?*

A. It is a tumor marker for choriocarcinoma.

47. *What is the mechanism of action of Mitomycin?*

A. Intercalation with DNA strands.

48. *What is the mechanism of action of Methotrexate?*

A. It is a folic acid antagonist.

49. What is vincristine and vinblastine?

A. They are alkaloids from vinca rosea, they interfere with assembly of cyto-skeleton and inhibit stathmokinesis (spindle movement), so they are used as anti-cancer drug.

50. What is the mechanism of action of adriamycin?

A. It inhibits topo-isomerase.

Chapter 24 *Complete the missed words*

Mention the vitamin deficiency of the following diseases:

1. Scurvy. . . . . .
2. Pellagra. . . . . .
3. Pernicious anemia: . . . . . .
4. Beri Beri. . . . . .
5. Osteomalcia. . . . . .
6. Night blindness. . . . . .

Mention the key enzyme (s) of following metabolic pathways

7. Heme synthesis. . . . . .
8. Bilirubin catabolism . . . . . .
9. Purine de novo synthesis . . . . . .
10. Lipolysis. . . . . .
11. Fatty acid synthesis (palmitate) . . . . . .
12. Urea synthesis. . . . . .
13. Cholesterol synthesis. . . . . .
14. Citric acid cycle . . . . . .
15. Glycogenesis . . . . . .
16. Glycogenolysis . . . . . .
17. Pentose phosphate pathway . . . . . .
18. Glycolysis. . . . . .
19. Gluconeogenesis. . . . . .

Mention one disease in which the following plasma enzymes are increased:

20.Gamma glutamyl transferase. . . . . .

21.ALT and AST. . . . . .

22.CK- AST and LDH. . . . . .

23.Amylase. . . . . .

24.Alkaline phosphatase. . . . . .

25.Acid phosphatase . . . . . .

26.Troponine C . . . . . .

27.Myoglobin . . . . . .

Mention the enzymatic defects of the following diseases:

28.Essential fructosuria . . . . . .

29.Hereditary fructose intolerance. . . . . .

30.Galactosemia . . . . . .

31.Lactose intolerance. . . . . .

32.Favism. . . . . .

33. Von Gierke's disease. . . . . .

34.Essential pentosuria . . . . . .

35.Steatorrhea due to pancreatitis. . . . . .

36.Tangier disease . . . . . .

37.Gaucher's disease . . . . . .

38.Niemann-Pick disease . . . . . .

39.Tay Sach's disease . . . . . .

40.Maple syrup urine disease. . . . . .

41.Albinism. . . . . .

42.Phenylketonuria. . . . . .

43.Alkaptonuria. . . . . .

44.Hyperammonemia type I . . . . . .

45.Hyperammonemia type II . . . . . .

46.Crigler – Najjar syndrome . . . . . .

47.Acute intermittent porphyria . . . . . .

48.Porphyria cutanea tarda. . . . . .

49.Hereditary coproporphyria. . . . . .

50.Variegate porphyria. . . . . .

51.Congenital erythropoietic porphyria . . . . . .

52. Protoporphyria . . . . . .
53. Succinyl choline apnea . . . . . .
54. Xeroderma pigmentosum . . . . . .

Mention the metabolic defects of the following diseases:

55. Diabetes mellitus. . . . . .
56. Lactic acidosis . . . . . .
57. Hypoglycemia. . . . . .
58. Ketosis. . . . . .
59. Kwashiorkor . . . . . .
60. Marasumus. . . . . .
61. Argintinofoma. . . . . .
62. Hartnup's disease . .. . . .
63. Glycinuria . . . . . .
64. Cystinuria . . . . . .
65. Myasthenia gravis. . . . . .
66. Prion disease . . . . . .
67. Alzheimer's disease . . . . . .
68. Parkinson's disease . . . . . .
69. Dubin – Johnson syndrome . . . . . .
70. Gelbert disease . . . . . .
71. hyperuricemia . . . . . .

Mention the metabolic disorder(s) resulting from deficiency of the following enzymes:

72. α-Ketoacid decarboxylase . . . . . .
73. Aldolase B . . . . . .
74. Dihydrobiopetrin reductase. . . . . .
75. Fructokinase . . . . . .
76. Galactose -1- phosphate uridyltransferase. . . . . .
77. Glucocerebrosidase . . . . . .
78. Glucose – 6 – phosphate dehydrogenase. . . . . .
79. Glucose -6- phosphatase. . . . . .
80. Histidinase . . . . . .
81. Lipoprotein lipase (LPL) . . . . . .

82. Pancreatic lipase . . . . . .
83. Para hydroxyphenyl pyruvate oxidase. . . . . .
84. Phenylalanine hydroxylase. . . . . .
85. Pyruvate kinase. . . . . .
86. Renal 1 α-hydroxylase. . . . . .
87. Sphingomyelinase . . . . . .
88. Tyrosine hydroxylase . . . . . .
89. Tyrosine transaminase . . . . . .
90. UDP-Glucuronyl transferase . . . . . .
91. UV specific endonuclease. . . . . .
92. Homogentisate oxidase. . . . . .
93. Carbamoyl phosphate synthase I. . . . . .

Mention the product(s) resulting from the reaction catalyzed by the following enzymes:

94. β-Carotene dioxygenase. . . . . .
95. Acetyl CoA carboxylase. . . . . .
96. Lecithin cholesterol acyltransferase (LCAT. . . . . .
97. Adenylate cyclase. . . . . .
98. ALA synthase. . . . . .
99. Alanine transaminase. . . . . .
100. Aldolase B. . . . . .
101. Citrate lyase. . . . . .
102. Dihydrofolate reductase . . . . . .
103. Glutaminase. . . . . .
104. Glyceraldehyde -3- phosphate dehydrogenase. . . . . .
105. Glycogen phosphrylase. . . . . .
106. Hepatic 25 cholecalceferol hydroxylase. . . . . .
107. HMG-CoA reductase. . . . . .
108. Hormone sensitive triacylglycerols lipase. . . . . .
109. L-Glutamate decarbxylase. . . . . .
110. Phenylalanine hydroxylase. . . . . .
111. Phosphfructokinase I. . . . . .
112. Phosphfructokinase II. . . . . .
113. Tyrosine hydroxylase. . . . . .

Mention the concentration of the following substances:

114. Fasting plasma glucose (mg/dl) . . . . . .
115. 2 Hours PP plasma glucose (mg/dl) . . . .
116. Total plasma cholesterol(mg/dl) . . . . . .
117. Total plasma calcium (mg/dl) . . . . . .
118. Indirect, unconjugated palsma bilirubin (mg/dl) . . . . ..
119. Direct, conjugated plasma bilirubin (mg/dl) . . . . . .
120. Plasma uric acid (mg/dl) . . . . . .
121. Plasma iron (mg/dl) . . . . . .
122. Plasma total iron binding capacity (mg/dl) . . . . . .
123. Plasma urea (mg/dl) . . . .
124. Urine urea (g/day) . . . .
125. Urine protein (mg/L) . . . . . .

Mention one organ / tissue location for the following compounds:

126. Collagen. . . . . ..
127. Elastin . . . . . .
128. Glycogen: . . . .
129. Heme. . . . . .
130. Immunoglobulin A. . . . . .
131. Keratin: . . . . . .
132. Myoglobin. . . . . .
133. Prostglandins. . . . . .

What are hydrolytic products of the following compounds:

134. Glutathione. . . . . .
135. Lctose. . . . . .
136. Sucrose. . . . . .
137. Maltose. . . . . .
138. Lecithin. . . . . .
139. ATP. . . . . .
140. NAD+. . . . . .

Answer Key

1. L-Ascorbic acid
2. Niacin
3. Vitamin B12
4. Vitamin B1
5. Vitamin D3 (calcitriol)
6. Retinal
7. ALA synthase
8. UDP-glucuronyl transferase
9. PRPP synthetase and PRPP glutamyl amidotransferase
10. Hormone sensitive triacylglycerol lipase
11. Acetyl CoA carboxylase
12. Carbamoyl synthase I
13. HMG CoA reductase
14. Citrate synthase, isocitrate dehydrogenase and alpha ketoglutrate dehydrogenase
15. Glycogen synthase
16. Phosphorylase
17. Glucose -6- phosphate dehydrogenase
18. Glucokinase, hexokinase, phosphofructokinase I, and Pyruvate kinase
19. Glucose -6- phosphatase, fructose 2,6 bisphosphatase, Pyruvate carboxylase, Phosphoenol pyruvate carboxykinase
20. Chronic alcoholism
21. Liver cell failure
22. Myocardial infarction
23. Pancreatitis
24. Obstructive jaundice
25. Prostatic carcinoma
26. Myocardial infarction
27. Myocardial infarction
28. Fructokinase
29. Aldolase B
30. Galactose -1- phosphate uridyl transferase, Galactokinase, UDP – Galactose epimerase.
31. Lactase

32.Glucose -6- phosphate dehydrogenase

33.Glucose -6- phosphate dehydrogenase

34.L-xylose reductase

35.lipase

36.LCAT

37.Beta glucosidase (glucocerebrosidase)...

38.Sphingomyelinase.

39.Hexosaminidase.

40.α-Ketoacid decarboxylase

41.Tyrosine hydroxylase

42.Phenylalanine hydroxylase

43.Homogentisic acid oxidase

44.Carbamoyl phosphate synthase I

45.Ornithin transcarbamoylase

46.UDP-Glucuronyl transferase

47.Uroporphyrinogen I synthase

48.Uroporphyrinogen decarboxylase

49.Coproporphyrin oxidase

50.Protoporphyrinogen oxidase.....

51.Uroporphyrinogen II synthase

52.Ferrochelatase

53.pseudocholinosterase

54.UV specific endonuclease

55.Insulin deficiency

56.Increased production of lactate or decrase utilization of lactate

57.Decrease blood glucose concentration below 45 mg/dl

58.Metabolic acidosis resulting from ketonemia.

59.hypoproteinosis results from deficiency of only dietary proteins in children

60.hypoproteinosis results from deficiency of dietary proteins, carbohydrate and lipds in children

61.Excessive production of serotonin by intestinal argentaffin cells on expense of niacin

62.Defective intestinal absorption and renal reabsorption of tryptophan.

63.dominant x-linked disease, due to defect in renal tubular reabsorption of glycycine

64. Defective renal tubular reabsorption of 4 amino acids cystine together with basic amino acids; lysine, arginine, and ornithine.
65. Antibodies against the nicotinic acetyl choline receptors that inhibit them.
66. A neuro degenerative disease due to aggregation and deposition of protein known as prion protein. It is insoluble protein.
67. It is a neurological disorder resulting from deposition a protein known as β-amyloid. It is insoluble protein.
68. Decrease of dopamine transmitter in mid brain.
69. Defect in the hepatic secretion of conjugated bilirubin into the bile.
70. Defect in the uptake of bilirubin by the liver cells
71. A condition in which serum urate level is increased above normal level (2-7 mg/dl) and exceeds its solubility limit.
72. Maple syrup urine disease
73. Hereditary fructose intolerance
74. Atypical phenylketonuria
75. Essential fructosuria
76. Galactosemia
77. Gaucher's disease
78. Favism
79. Von Gierke's diseas
80. Histidinemia
81. Primary hyperlipoproteinemia
82. Steatorrea
83. Tyrosinemia
84. phenylketonuria
85. Hemolytic anemia
86. Renal rickets
87. Niemann-Pick disease
88. Albinism
89. Tyrosinemia
90. Crigler Najjar syndrome
91. Xeroderma pigmentosum
92. Alkaptonuria
93. Hyperammonemia type I
94. Retinal
95. Malonyl CoA

96. Cholesterol ester + Lysolecithin
97. Cyclic AMP
98. Delt amino levulonic acid
99. Pyruvate + Glutamate
100. Dihydroxyacetone phosphate + Glyceraldehyde
101. Acetyl CoA + Oxaloacetate
102. Tetra hydrofolate
103. Glutamate + Ammonia
104. 1,3 Bisphosphoglycerate
105. Glucose -1- phosphate
106. 25 cholecalceferol
107. Mevalonate
108. Diacylglycerol + Fatty acid
109. Gamma amino butyric acid (GABA)
110. Tyrosine
111. Fructose 1,6 bisphosphate
112. Fructose 2,6 bisphosphate
113. DOPA
114. 65 -110 mg/dl
115. 65 -140 mg/dl
116. 140 - 220 mg/dl
117. 8.5 -10.5 mg/dl
118. 0.2 – 1.0 mg/dl
119. 0.0 – 0.2 mg/dl
120. 2 – 7 mg/dl
121. 60 – 160 mg/dl
122. 180 – 450 mg/dl
123. 20 - 50 mg/dl
124. 20 - 40 g/day
125. Less than 30 mg/L
126. Tendons and bones
127. Lung and blood vessels
128. Liver and muscles
129. RBCs and muscles
130. Body secretions
131. Skin, hair and skin

132. Skletal muscles
133. Seminal fluids
134. Glutamate, cysteine and glycine
135. Glucose and galactose
136. Glucose and fructose
137. Glucose and glucose
138. Glycerol + saturated fatty acid + unsaturated fatty acid + phosphate + choline
139. Adenine + Ribose + phosphate
140. Nicotinamide + Ribose + phosphate + Adenine + Ribose + phosphate

CHAPTER 25 SHORT CASES IN CLINICAL BIOCHEMISTRY

1. The following plasma biochemical profile was obtained from a semicomatosed patient admitted to the Al-Azhar University Hospital:

Test	Value		Reference range	
Glucose, fasting	700	mg/dl	65 - 110	mg/dl
Cholesterol	300	mg/dl	< 220	mg/dl
Ketone bodies	100	mg/dl	1 - 3	mg/dl
Potassium	5.5	mmol/L	3-5 - 5	mmol/L

A. On biochemical bases what is the provisional diagnosis?

B. Explain accordingly, why these metabolites and electrolyte are elevated.

C. During the treatment, explain why potassium should be given?

2. A 45-year old diabetic woman was admitted semicomatosed to the hospital. She has headache, confusion, anxiety, profuse sweating and hunger. On examination she was hypotensive and has tachycardia and tremors of hands.

A. What is the provisional diagnosis?

B. What are the possible causes?

C. What are the investigations required to be done immediately.

3. A 79-year old woman was admitted to the hospital deeply unconscious and extremely dehydrated. No history was available. The admitting doctor arranged some plasma biochemical results, which were as follows:

Test	Value		Reference range	
Glucose	775	mg/dl	65 - 110	mg/dl
Ketone bodies	1.9	mg/dl	1 - 3	mg/dl
Urea	132	mg/dl	20-50	mg/dl

Creatinine	1.4	mg/dl	0.7-1.2	mg/dl
Biocarbonate	23	mmol/L	21-25	mmol/L
Cl	120	mmol/L	96-106	mmol/L
Na^+	155	mmol/L	134-147	mmol/L
K^+	5.1	mmol/L	3.5 - 5	mmol/L
pH	7.39		7.35-7.42	

A. What is the provisional diagnosis?

B. What are actions needed?

4. A 9 months baby was presented to the hospital with jaundice. His mother reported that the baby was normal until he has feed a little smashed fava bean; at that time he began to experience fever and shivering. His skin turned yellow and his urine turned dark brown.

A. What is the provisional diagnosis?

B. What is the possible biochemical defect?

C. What are investigations required?

D. What are recommendations you advise the child's parents.

5. *A 3 years old child was referred to pediatric emergency room, suffering of dizziness, confusion, loss of concentration, anxiety, profuse sweating and hunger. On examination he was found to have enlarged liver. Liver biopsy was taken and proved to contain excess glycogen than normal.*

The admitting doctor arranged some plasma biochemical results, which were as follows:

Test	Value		Reference range	
Glucose	42	mg/dl	65 - 110	mg/dl
Lactate	3.2	mmol/L	1 - 2	mmol/L
Ketone bodies	9.2	mg/dl	1 - 3	mg/dl
AST	63	UII	10-37	U/L
ALT	55	UII	10-42	U/L
Triacylglycerols	180	mmol/L	40-160	mg/dl
Uric acid	8.4	mg/dl	2-7	mg/dl

A. On biochemical bases what is the provisional diagnosis?

B. What is the possible biochemical defect?

C. Explain accordingly, why these metabolites are changed?

6. A 22 years old female attended an internal medicine clinic, complaining of diarrhea, distention, and abdominal cramps. She gave history of ingestion large amount of milk. On examination she showed signs of dehydration.

   A. *What is the provisional diagnosis?*

   B. *What are the possible biochemical defects?*

   C. *What are mechanisms of these signs and symptoms?*

7. A 3 years old hyperactive baby was referred to pediatrician, suffering from mental retardation, tremors, failure to walk and talk. His IQ is below 50 (normal 90-110). Blood was drawn for amino acids analysis. Among them, phenylalanine was proved elevated.

   A. *What is the provisional diagnosis?*

   B. *What are the possible biochemical defects?*

   C. *How can these conditions be prevented?*

8. A 45 years old male patient referred to an emergency room, suffering of dizziness, confusion, loss of concentration, blurring vision and vomiting. On examination he was found to have flapping tremors. The following liver function tests were done:

Test	Value		Reference range	
Ammonia	890	Umol/l	10-110	Umol/l
Total bilirubin	27	mg/dl	0.2-1.2	mg/dl
Direct bilirubin	22	mg/dl	0.0-0.2	mg/dl
AST	2650	U/L	10-37	U/L
ALT	3481	U/L	10-42	U/L
Alkaline phosphatase	932	U/L	100-290	U/L

   A. *What is the provisional diagnosis?*

   B. *What are the possible causes of this condition?*

   C. *What are is the mechanism of these symptoms and signs?*

   D. *Explain the urgent treatment needed*

9. A 6 months baby presented with jaundice, diarrhea, vomiting and failure to grow. Plasma amino acids showed marked elevation of tyrosine. Liver function tests were elevated. The baby was died one month later from liver cell failure.

A. *What is the provisional diagnosis?*

B. *What is the possible biochemical defect?*

C. *How can this condition be prevented?*

10. *A 5 years old child was referred to pediatrician, suffering from arthritis. His urine turned black when left to stand.* Plasma homogentisate showed marked elevation.

A. *What is the provisional diagnosis?*

B. *What are the possible biochemical defects?*

11. A middle aged man with a family history of ischemic heart disease had fasting lipids studies performed, with the following results:

Test	Value	Reference range	
Cholesterol	198	< 220	*mg/dl*
Triacylglycerols	347	*60 - 160*	*mg/dl*
Lipoprotein electrophoresis	Increased pre-beta fraction		

A. *What is the lipid disorder,*

B. *What are the possible causes of this condition?*

12. A 43 years old man attended orthopedic clinic sffuring from acute attack of severe pain in the big toe, which was swollen and red.The patient gave history of renal stones.

A. *What is the provisional diagnosis?*

B. *What are the possible biochemical defects?*

C. *How can these conditions be treated?*

13. A 60-year old man developed jaundice associated with weight loss but no pain. Physical examination was normal except there was deep icterus. The liver function tests were as follows:

Test	Value		Reference range	
Bilirubin (total)	13	mg/dl	0.1-1.2	mg/dl
Bilirubi (direct)	12	mg/dl	0.0-0.2	mg/dl
ALT	25	U/l	10-37	U/L
AST	19	U/l	10-42	U/L
Alkaline phosphatase	570	U/l	100-290	U/L

A. *What type of jaundice is this?*

B. *What is the likely diagnosis?*

14. A 58-year old man became unwell and developed jaundice. His doctor arranged for some liver function tests (LFTs), which were:

Test	Value		Reference range	
Bilirubin (total)	3.4	mg/dl	0.1-1.2	mg/dl
Bilirubi (direct)	2.1	mg/dl	0.0-0.2	mg/dl
ALT	180	U/l	10-37	U/L
AST	157	U/l	10-42	U/L
Alkaline phosphatase	340	U/l	100-290	U/L

A. On biochemical bases what is the provisional diagnosis?

B. Explain accordingly, why these metabolites and enzymes are elevated.

C. What are the possible causes?

15. A 4 days old newborn presented in neonatology clinic with jaundice. The following biochemical tests were done:

Test	Value		Reference range	
Bilirubin (total)	8.5	mg/dl	0.1-1.2	mg/dl
Bilirubi (direct)	0.3	mg/dl	0.0-0.2	mg/dl
ALT	16	U/l	10-37	U/L
AST	18	U/l	10-42	U/L
Alkaline phosphatase	110	U/l	100-290	U/L

A. What is the provisional diagnosis?

B. Explain accordingly, why total bilirubin is elevated.

C. What is the effect of elevated bilirubin?

D. How to manage the case?

16. A 37-year old man did a preoperative examinations and investigations. The physician noted that the patient was asymptomatic and had no abnormal physical signs. The liver function tests were normal except for total bilirubin, 2.9 mg/dl (normal 0.2-1.2 mg/dl). The operation was postponed. The physician asked to repeat the test for bilirubin after 24 hours caloric restriction. Total bilirubin was elevated to 4.2 mg/dl.

A. What is the most likely diagnosis?

B. What type of hyperbilirubinemia is this?

17. A 44-year old farmer was adimiited with diarrhea, dermatitis in the exposed area of skin and dementia. His family gave dietary history mainly of corn and corn products.

A. What is the most likely diagnosis?

B. Explain the metabolic causes of these signs.

C. What will be the treatment?

18. 35-year old female presents to the clinic feeling tired, fatigued and loss of interest in normal daily activities. She has problem of constipation, muscle cramps, loss of interest in sex, and menstrual disorders. She is frequently cold when others are hot. Her skin has become dry, and she has noticed a swelling sensation in her neck area. On the examination she is febrile with a slower heart rate pulse 60/min. She has an enlarged, non tender thyroid noted on her neck. Her reflexes are diminished.

A. What is the most likely diagnosis?

B. What laboratory test would you need to confirm the diagnosis?

C. What are the metabolic changes in the patient?

19. A 3-year old boy is brought to the emergency department after several episodes of vomiting and lethargy. His pediatrician has been concerned about his failure to thrive and possible hepatic failure along with recurrent episodes of the vomiting. After a careful history is taken, you observe that these episodes occur after ingestion of certain types of food, especially high in fructose. A blood sugar was checked and was extremely low.

A. What is the most likely diagnosis?

B. What is the biochemical basis for clinical symptoms?

C. What is the treatment of the disorder?

20. A 50-year old female presents to your clinic with complaints of excessive thirst, fluid intake, and polyurea. She denies any urinary tract infection symptoms. She reports no medical problems, but has not seen a doctor in many years. On examination she is an obese female in no acute distress. Her physical examination is otherwise normal. The urine analysis revealed large glucose, and a serum random blood sugar level was 320 mg/dl.

A. What is the most likely diagnosis?

B. What are the causes of metabolic changes in the patient?

C. What is the biochemical basis of this disease?

21. A 30 year old male presents with evidence of myocardial infarction (severe retro-sternal pain which is referred to left shoulder).

A. ***What are the possible biochemical tests you asked for to confirm the diagnosis?***

B. ***When asked for each test?***

Answers

1. **Case taking number 1:**
   A. The provisional diagnosis is severe UNCONTROLLED DIABETES MELLITUS.
   B. Causes of elevated metabolites and electrolytes: Insulin deficiency causes: hyperglycemia (700 mg/dl). Insulin deficiency also causes excessive lipolysis ➔ high acetyl CoA ➔ hypercholesterolemia and excess ketone bodies. Increased plasma potassium is due to mobilization of potassium from inside to outside the cells associated with insulin deficiency.
   C. Potassium should be given because insulin treatment will cause potassium to go inside cells with subsequent hypokalemia.

2. **Case taking number 2:**
   A. The provisional diagnosis is HYPOGLYCEMIA.
   B. The possible causes are either taking insulin treatment without food intake or excessive dose of insulin injection.
   C. Testing blood glucose.

3. **Case taking number 3:**
   A. The provisional diagnosis is HYPEROSMOLAR NON-KETOTIC DIABETIC COMA.
   B. This is a hyperglycemic diabetic emergency, and the normal plasma ketone bodies, CO_2 do not suggest ketosis. There is also hypernatremia and hyperosmolality confirming hyperosmolar non-ketotic diabetic coma.
   C. Actions needed: This patient carefully rehydrated with half-normal saline and given low dose insulin infusion.

4. **Case taking number 4:**
   A. The provisional diagnosis is FAVISM.
   B. The possible biochemical defect is the deficiency of glucose-6-phosphate dehydrogenase enzyme.
   C. The investigations required are estimation of red cells glucose-6-phosphate dehydrogenase enzyme, reticulocytes, hemoglobin concentration, and estimation of plasma total and direct bilirubin,

D. The recommendations for the child's parents are to avoid ingestion of fava beans, antimalarial drugs and some antibiotics as streptomycin.

5. Case taking number 5:

A. The provisional diagnosis is VON GIERKE'S DISEASE.

B. The possible biochemical defect is the hereditary deficiency of glucose-6-phosphatase enzyme.

C. Deficiency of the enzyme prevent glycogenolysis i.e. conversion of glycogen into glucose. This leads to:

1. Accumulation of large amount of glycogen in liver. This leads to **disturbance of liver functions ➔ Elevated ALT and AST.**
2. **Enlargment of liver** (hepatomegally).
3. **Fasting hypoglycemia.**
4. **Ketosis** and hyperlipidemia as lypolysis and fatty acid acids oxidation increased to give energy required ➔ ↑ Ketone bodies..
5. **Hyperuricemia (gout):** ↓ glucose-6-phosphatase ➔ ↑ Glucose-6-phosphate ➔ ↑ Pentose phosphate pathway ➔ ↑ Ribose production ➔ ↑ Uric acid ➔ Gout.

6. Case taking number 6:

A. The provisional diagnosis is LACTOSE INTOLERANCE.

B. The possible biochemical defect is the acquired deficiency of intestinal lactase, essential for digestion of lactose.

C. Mechanism of signs and symptoms: The presence of lactose in intestine causes:

1. *Increased osmotic pressure: So water will be drawn from the tissue (causing dehydration) into the large intestine (causing diarrhea).*
2. *Increased fermentation of lactose by bacteria: Intestinal bacteria ferment lactose with subsequent production of CO_2 gas. This causes distention and abdominal cramps.*

7. ***Case taking number 7:***

A. The provisional diagnosis is PHENYLKETONURIA.

B. The possible biochemical defect is the hereditary deficiency of phenyl alanine hydroxylase enzyme.

C. Phenylketonuria can be prevented by:

1. **Diagnosis:** All newly born infants are screened at birth by measuring blood phenylalanine. Abnormal high phenylalanine level will be found in cases of phenylketonuria.
2. Any infant proved to have abnormal high level of blood phenylalanine, should feed **milk containing very low amount of phenylalanine.**
3. This regimen of diet is maintained up to 6 years of age when a high concentration of phenylalanine has no longer effect on brain cells.

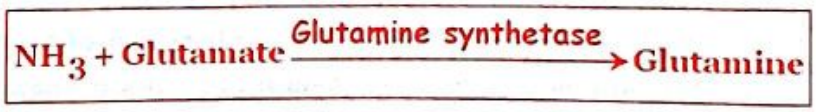
8. ***Case taking number 8:***

A. The provisional diagnosis is HYPERAMMONEMIA due to liver cell failure.

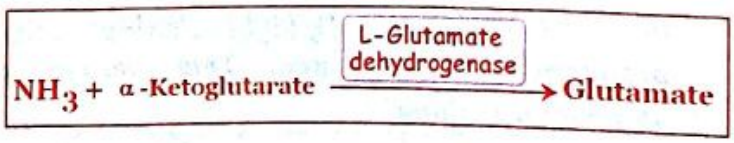
B. The possible causes of this condition are acquired liver disorders as advanced liver fibrosis and cirrhosis.

C. Mechanism of symptoms and signs:

1. At normal blood ammonia level , any ammonia reaches the brain is incorporated into glutamine formation by glutamine synthetase enzyme.



2. In cases of hyperammonemia , ammonia reacts not only with glutamate , but also with α-ketoglutarate by glutamate dehydrogenase enzyme. This depletes α-ketoglutarate which is an essential intermediate of citric acid cycle ➔ Decrease



in ATP and energy production ➔ symptoms of ammonia intoxication and even coma.

D. The urgent treatment needed is giving glutamate infusion to save α–ketoglutarate.

9. Case taking number 9:

A. The provisional diagnosis is HEREDITARY TYROSINEMIA (TYROSINOSIS):

B. The possible biochemical defect is the hereditary deficiency of either tyrosine α-ketoglutarate transaminase or p-hydroxy-phenylpyruvate oxidase enzymes essential for tyrosine metabolism.

C. Tyrosinosis can be prevented by feeding the affected infant and children a diet containing very low levels of tyrosine, methionine and phenylalanine (precursor of tyrosine).

10. Case taking number 10:

A. The provisional diagnosis is ALKAPTONURIA.

B. The possible biochemical defect is a hereditary deficiency of homogentisic acid oxidase.

11. Case taking number 11:

A. The lipid disorder is Type IV hyperlipproteinemia. Hypertriacylglerolemia with a raised pre-beta lipoprotein represents a Fredericksen Type IV abnormality.

B. *The possible causes* may be secondary to other conditions such as alcoholism, obesity, diabetes, pancreatitis, chronic renal failure, acromegally and gout. It may also occur as a familial disease. In this case the patient was obviously overweight, and lipids fell to normal on successful dieting.

12. Case taking number 12:

A. The provisional diagnosis is GOUTY ARTHRITIS.

B. The possible biochemical defects?

1. **Primary hyperuricemia (enzyme defects):** ↑ PRPP synthase, ↓ HGPRTase (Lesch nyhan syndrome) and ↓ glucose-6-phosphatase (Von Gierke's disease).
2. **Secondary hyperuricemia:** ↑ The rate of cell division and tissue turnover and ↓ Renal excretion of uric acid.

C. Treatment of these conditions:

- Treatment of the cause.
- **Allopurinol:** It is a structural analogue of hypoxanthine that competitively inhibits xanthine oxidase enzyme → decreasing formation of uric acid.

13. ***Case taking number 13:***

A. The provisional diagnosis is **OBSTRUCTIVE JAUNDICE**.

B. The diagnosis is based on the liver function tests (LFTs) which indicate that the type of jaundice is obstructive jaundice. This is due to marked elevation of plasma bilirubin and most of it is of direct type. Other liver function tests indicate high concentration of alkaline phosphatase and normal amino transferases concentration. Obstructive jaundice is nearly due to either gall stones or malignancy (cancer head of pancreas). The age of the patient and history of loss of weight suggests obstructive jaundice due to cancer head of pancreas.

14. ***Case taking number 14:***

A. The provisional diagnosis is **HEPATOCELLULAR JAUNDICE** as indicated by The LFTs.

B. In hepatocellular jaundice there a failure in liver functions as well as some sort of obstruction. This is manifested by a moderate elevation of direct and indirect bilirubin. Elevated ALT and AST indicate destruction of liver cells. Elevated alkaline phosphatase indicates some sort of obstruction.

C. The possible causes may be bilharziasis and viral hepatitis (B &C). Hepatitis A is unusual in patients in this age. Tests for hepatitis B and C are recommended. Bilharzial test is also recommended to exclude bilharzial cirrhosis.

15. ***Case taking number 15:***

A. The provisional diagnosis is **PHYSIOLOGIC (NEONATAL) JAUNDICE** as indicated by The LFTs.

B. **Why total bilirubin is elevated.** At birth, liver contains very little UDP-glucuronyl-transferase enzyme, which is important for conjugation of bilirubin. Also at birth, there is an accelerated hemolysis of RBCs. This leads to increased unconjugated bilirubin and jaundice.

C. **Effect of elevated bilirubin:** If unconjugated bilirubin exceeds the concentration, which can be tightly bound to plasma albumin (20-25 mg/dl), free bilirubin can pass blood brain barrier, causing damage to the brain centers of infants. This is called Kernicterus. It may cause mental retardation.

D. How to manage the case? Neonatal jaundice is treated by phenobarbital and exposure of jaundiced baby to ultraviolet light (photo- therapy) as bilirubin is broken down in light.

16. **Case taking number 16:**

A. The most likely diagnosis is GILBERT SYNDROME as indicated by normal liver function tests and absence of symptoms. Also bilirubin gets increased by caloric restriction.

B. Type of hyperbilirubinemia: unconjugated as the defect is mainly in the uptake of bilirubin by the liver.

17. **Case taking number 17:**

A. The most likely diagnosis is NIACIN DEFICIENCY (PELLAGRA).

B. The metabolic causes are dietary deficiency of niacin, tryptophan and/or vitamin B_6 essential for niacin formation. Corn is deficient in niacin and tryptophan.

C. Treatment include diet regimen which formed of niacin rich food. Oral nicotinamide is recommended.

18. **Case taking number 18:**

A. The most likely diagnosis is HYPOTHYROIDISM.

B. The laboratory tests needed to confirm the diagnosis are thyroid function tests mainly measuring T3, T4 and TSH.

C. The metabolic changes in the patient: Thyroid hormones increase heat production and oxygen consumption in most tissues through stimulation of ATPase activity. They act together with growth hormone as major anabolic agents during growth. They stimulate DNA in the nucleus of cells. Deficiency of thyroid hormones causes many of the body's functions to slow down ➔ Signs and symptoms of hypothyroidism.

19. **Case taking number 19:**

A. *The most likely diagnosis is* HEREDITARY FRUCTOSE INTOLERANCE.

B. *The biochemical basis for clinical symptoms:* the accumulation of fructose-1-phosphate leads to:

1. Damage of liver and kidney tissues ➔ Liver and kidney failure.

2. Inhibition of phosphorylase enzyme. This leads to inhibition of glycogenolysis and fasting hypoglycemia.

C. *The treatment of the disorder is avoidance of fruit juice, fruits and sweets. Fructose low diet is generally recommended.*

20. Case taking number 20:

A. *The most likely diagnosis is* TYPE II DIABETES MELLITUS.

B. *The cause of biochemical basis of this disease is the deficiency of insulin hormone which disturbs carbohydrate, lipids, protein, minerals and vitamins metabolism.*

C. *The metabolic changes in the patient are due to* insulin deficiency which leads to decrease glucose uptake by tissues, ↓ glucose oxidation, ↑ gluconeogenesis and ↑ Glycogenolysis. This results to:

1. ↑ Blood glucose (hyperglycemia), this causes ↑ Plasma osmolality ➔ dehydration of body cells → Sense of thirst.
2. Glycosuria: If blood glucose level exceeds renal threshold ➔ Glucose appears in urine (glycosuria). This will leads to ↑ osmotic diuresis ➔ Excessive and frequent urination (polyurea).

21. Case taking number 21:

A. Ask for biochemical tests which include: CK, CK-MB, AST, LDH and cardiac troponin I.

B. CK level starts to rise within three hours of infarction. It can be used for diagnosis in the first 24 hours. AST can be used 2-3 days after onset of infarction. LDH can be used 5-7 days after infarction. Cardiac troponin I (CTI) is released into the blood within four hours after the onset of cardiac symptoms, peaks at 12-16 hours and remains elevated for 5-9 days post-infarction. Therefore, CTI is very useful as a marker at any time interval after the heart attack. It is 75% sensitive index for myocardial infarction.

Biochemistry

Questions and Answers

This book "Oraby's Biochemistry " by SAID ORABY is made in it's four parts (I, II, III and IV) to provide necessary knowledge and recent information about biochemistry for medical students and allied sciences.

- All efforts have been made to simplify most of the subjects.
- Latest advances in biochemistry important to medicine.
- Many illustrations are added to bring biochemistry alive.
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- Postgraduates and students who are preparing for standard courses or examinations (fellowships, ECFMG.. etc) will find this book of benefit for them.
- Finally, I hope this work is appreciated and accepted by students and colleagues.

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